DISSERTATION ON

A STUDY ON LIMB SALVAGE IN EXTREMITY SOFT TISSUE SARCOMAS

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BONAFIDE CERTIFICATE

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DISSERTATION ON
SOFT TISUE SARCOMA

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CONTENTS

INTRODUCTION

INTRODUCTION

Extremity sarcomas are rare(less than 1% of all malignancies) and pose a challenging problem for patients and their surgeons / oncologist. In past, these cancers have treated by amputation with relatively poor functional outcome. Over the last 30 yrs , limb salvage has evolved , this technique has been proven safe and effective . There have been developments in several areas to improve the outcomes with limb preservation. The neoadjuvant chemotherapy causes tumour necrosis, which allows for safer removal. In addition Chemotherapy causes the tumour to develop a rind or margin and in some cases Shrink , helping the surgeon to completely resect the tumour and minimize the removal of normal tissue. Imaging also play a major role in limb salvage with development of radiographic studies such as MRI .The surgeon can better see the extent of tumour and plan resection precisely.

Finally, there have been major development in limb reconstruction. Surgeons now have available implants that can be matched to the patients size, stronger metals, bone and soft tissues allografts micro vascular muscular transfer procedure. These techniques allow the limb to be resorted with good function.

Despite all of these improvement and enthusiasm for limb salvage , It is not For everyone . There are contraindications and complications that need to be considered. The decision needs to have a thorough understanding of the advantages and disadvantages of limb salvage before pursuing this technique.

AIM OF THE STUDY

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This is the era of minimal surgery and maximal functional outcome without compromising oncological principles and survival benefit. Before 1990, nearly 90% of the extremity soft tissue sarcomas were treated by amputation. But after 1990, about 90% of these tumours are treated by limb sparing surgeries, thanks to better understanding the nature of soft tissue tumours, and improvement in micro vascular techniques and development in prosthetic field.

This is a retrospective analytical study about extremity soft tissue sarcomas in our institution for past 5years (2001-2005). This study thoroughly analyze the epidemiology, anatomical distribution , common histological types of soft tissue sarcomas in extremities . But main focus of this study is , to analyze the proportion of patients underwent limb sparing surgeries, on what basis patients were offered either limb sparing procedure or amputations . Since our institute is tertiary centre many soft tissue sarcomas are referred with local recurrence due to unplanned first surgery, this study mainly analyze the feasibility of limb salvage treatment in these patients and the factors adversely affect treatment outcome.

This study to analyze about local recurrence after multimodality treatment especially after limb sparing procedure whether these procedures are compromising the oncological principle and affecting overall survival rate . And to analyze about impact of multimodality therapy in saving the patient's limb. Finally, this study analyze about whether achieving local control has any consequence in development of distant metastasis.

LITERATURE REVIEW

HISTORICAL REVIEW

PAPYRUS EBERS described cancer as a tumour which is not to be touched as early as 1552BC. HIPPOCRATUS (460-375 BC) believed that cancer is incurable and described about nasal angiofibroma . GALAN, ALBUCASIS and later ARDRIUE (1306AD) refrained from surgery and were pessimistic. With advent of microscope KOCHER (1602-1680), LEUWENHOEK (1632-1732) gave description of cell that caused diseases. In 1712, EHNULLERUS published a book on of tumours .The term 'soft cancers'was introduced by WARDROP(1782-1869)

The credit of differentiating soft cancer from carcinomas must be given to CHARLESBELL (1774-1842) who first described and illustrated soft cancer of thigh in 1816. ABERNETHY (1780-1848) gave first classification of sarcomas in 1811 . MALHNEY in 1866 wrote the first monograph on fibrous tumours, Between 1860-1870, most of sarcomas were identified . ADAMS IN 1898 , BORST IN 1902 classified connective tissue tumour more extensively. In 1910, HARRISION introduced tissue culture technique and ROVES in 1911 transferred sarcoma to a chicken by inoculating cell free infiltrate.

In classification and morphology of STS the credit belongs to A.P.STOUT who attempted to lay the foundation for classification and sub classification of STS as early as 1947.

ETIOLOGY

GENETIC

Involvement of genetic factors in the genesis of soft tissue sarcoma is manifest by strong hereditary tendency for certain sarcomas (Strong,1977; Littlefield, 1984; Li et al, 1988; Rowley, 1988). Gardner's syndrome is an hereditary disease, one feature of which is desmoid tumours (McAdam and Goligher,1970); neurofibromatosis I also features tumour of the soft tissues: neurofirbromas and neurofibrosarcoma. A significant proportion of these patients ultimately exhibit transformation of the neurofibroma into neurofibrosarcoma (Fraumeni, 1973; Strong, 1977; Zoller et al, 1977). This disorder is associated with a mutation in the NF1 gene. It has been proposed that malignant degeneration reflects the two-hit hypothesis in which one allele is constitutionally inactivated in the germline while the other allele undergoes somatic inactivation (the second hit)(Colman et al., 1955). Sarcomas of soft tissue and bone, particularly osteosarcoma, have been observed later in life in surviving patients with familial or bilateral retinoblastoma (Derkinderen et al., 1988 Wong et al., 1997). Patients with Li-Fraumeni syndrome often develop sarcomas (Li-Fraumeni , 1969; Li et al., 1988). The Li-Fraumeni syndrome is inherited as an autosomal recessive trait, and is primarily characterized by soft tissue and bone sarcomas and breast cancer; other features include brain tumour, leukemia and adrenocortical cancer occurring before the age of 45 (Li and Fraumeni, 1969; Li et al ., 1988). Some patients develop multiple malignancies (Hisada et al., 1998). A germline mutation in

the p53 tumour suppressor gene is found in most affected families (Malkin, 1993; Evans and Lozano, 1997; Varley et al, 1997).

MOLECULAR BIOLOGY

Deletions or mutations of the tumour suppressor retinoblastoma (Rb) gene are critical in the pathogenesis of retinoblastoma and a variety of solid tumours. Alterations in the Rb gene are common in soft tissue sarcoma (Cance et al., 1990; Wunder et al., 1991; Karpeh et al., 1995), occurring in up to 70 percent of tumours (Cance et al., 1990; Karpeh et al., 1995). It has been proposed that Rb alterations are primary events in human sarcomas and many be involved in tumorigenesis or the early phases of tumour progression Karpeh et al., 1995). The Rb gene is critical for proper entry and transition through cell cycle. This change has been shown to be associated with infrequent osteogenic or other sarcomas as second malignant neoplasm in patients with hereditary retinoblastoma (Friend et al., 1987). Furthermore, the absence of detectable RB protein is associated with a small proportion of apparently sporadic sarcomas (Shew et al., 1989). In an examination of 43 sarcomas of bone and soft tissues, Wunder et al., (1991) found alteration in the RB gene in approximately 40 percent of the tumours.

The gene p53 is also a 'tumour suppressor gene' and mutation on this gene and/or absence of its protein is associated with the development of sarcomas and other malignancies. The p53 protein is a transcriptional activator that plays a key role in the integration of signal; inducing cell division, arrest of DNA synthesis following DNA damage, and programmed cell death (apoptosis). DNA damage results in increased

levels of p53 protein which induces cell cycle arrest at the G1/S interface, thereby permitting the cell to repair genomic damage or to initiated apoptosis .

Somatic mutations in the p53 gene are the most frequently detected molecular alteration in sporadic soft tissue sarcoma. These mutations have been detected in a variety of soft tissue sarcomas including malignant fibrous histiocytoma(MFH), leiomyosarcoma, liposarcoma, and rhabdomyosarcoma .

The MDM2 gene, located at 12q13-14, is overexpressed in a variety of human tunmouirs including soft tissue sarcomas (Oliner et al., 1992, khatuv et ak., 1993; Florenes et al., 1994; Nilbert et al., 1994). Its gene product localizes predominantly to the nucleus, where it acts as an inhibitor of the p53 tumour suppressor gene product. The gene SAS has been observed to be amplified in a high proportion of malignant fibrohistocytomas and liposarcomas (Smith et al., 1992). Furthermore, (uda et al., 1993) reported that approximately 40 percent of sarcomas were found to have an increased expression of c-erb-B2 oncogene and epidermal growth factor receptor (EGFR). Additional gene mutations or other alterations in sarcomas include: CDK4 (Kanoe et al., 1998), nrs, myc, and c-fas (Brown et al., 1984; Chardin et al., 1985).

CYTOGENETICS

Most synovial cell sarcomas are characterized by the translocation t(x;18)(p11.2q11.2). The breakpoint of this translocation fuses the SVT gene from chromosome 18 to one of two homologous genes, SSX1 OR SSX2 on the X chromosome (Clark et al., 1994; Kawai et al., 1998). The SVT-SSX gene is thought

to function as an aberrant transcriptional regulator. The nature of the chimeric gene appears to have prognostic and pathogenetic importance, as metastasis-free survival is much higher (relative risk 3.0) with SVT-SSX2 compared to SVT-SSX1 (Kawau et ak., 1994). SVT-SSX1 is associated with biphasic tumours (glandular epithelial differentiation on a background of spindle tumour cells), while SVT-SSX2 us associated with monophasic tumours that lack glandular epithelial differentiation.

Other chromosomal changes characteristic of specific sarcoma type include the reciprocal exchange t(1;22) (q24;q12) seen in approximately 85-90 percent of Ewing's sarcoma and primitive neuroectodermal tumours (PNET) (Turc-Carel et al., 1988; Landanyl et al., 1990; Delattre et al., 1994). In this translocation, the EWS gene from chromosome 22q12 is covalently linked to the ETS family member, FLI -1 (Zucman et al., 1993). The chimeric proteins that result from this translocation may alter transcription of an unidentified gene on chromosome 22 (Delattre et al., 1992; Zucman et al., 1993). A less common translocation t(21;22)(a22;q12) has also been identified and links to EWS to a different ETS family member, ERG (Sorensen et al., 1994).

Myxoid and round cell subtypes of liposarcomas display a reciprocal translocation t(12;16)(q13;p11) (Crozat et al., 1993; Rabbitts et al., 1993). In this translocation, the CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone; induced by DNA damage) gene is inserted adjacent to a novel gene called FUS. The fusion gene, called TLS-CHOP, shows sequence homology to the Ewing's fusion gene (Aman et al., 1992; Crizat et al., 1993; Rabbitts et al., 1993; Hisaoka et al., 1998). It falls to

induce G_1/S arrest, which is one of the functions of the non-oncogenic form of CHOP (GADD153)(Barone et al., 1994). Identification of the fusion gene has been used as a diagnostic aid for these subtype of liposarcoma (Hisaoka et al.,1998).

Alveolar rhabdomyosarcomas show a translocation at t(2;13)(q35;q14) or less often t(1;13)(p36;q14); the chimeric genes have been cloned and have been termed PAX3-FKHR and PAX7-FKHR, respectively (Davis and Barr, 1997). PAX7-FKHR tumours more often present with extremity lesions, are more likely to be localized, and are less likely to metastasize widely than PAX3-FKHR tumours (Kelly et al., 1996b). A downstream target of PAX3-FKHR may be MET, which encodes a receptor involved in growth and motility signaling (Ginsberg et al., 1998).

Clear cell sarcoma is usually classified as a malignant melanoma, although cytogenetically the tumours are distinct. Clear cell sarcomas often exhibit a translocation at t(12;22)(q13-14;q12), which is not seen in malignant melanoma (Fletcher, 1992). Trisomy of chromosome 8 is also observed in clear cell sarcoma (Travis and Bridge, 1992). Alterations in the 12q13-15 region has been described in a subgroup of haemangiopericytomas (Sreekantaiah et al., 1991; Mandahl et al., 1993). Multiple chromosomal abnormalities have also been seen in some of these tumours (Sreekantaiah et al., 1991).

ENVIRONMENTAL FACTORS

Radiation is recognized as capable of inducing sarcoma of bone and soft tissue. The frequency increases with radiation dose and with the post-radiation observation period (Kim, 1978; Sadove et al., 1981; Robinson et al., 1988). The most frequent histopathological type of radiation-induced sarcoma arising in soft tissues is malignant fibrous histiocytoma (approximately 70 percent).Although seen rarely after low doses (<40 Gy), this is predominantly a complication of high-dose treatment. The actuarial frequency at 15-20 years is approximately 0.5 percent for radiation of normal bone and soft tissue in the adult treated with radiation alone to full dose.

The frequency is higher following treatment of children, especially with radiation and chemotherapy; the frequency may be as high as 20-30 percent at remote times .

Chemotherapeutic agents are likewise associated with risks of sarcoma induction. For example, there are two reports that describe the appearance of osteosarcoma in children treated for leukaemia by drugs alone (Shaw et al., 1988). Tucker et al, (1987) analyzed the late sequels in 9170 long-time survivors of childhood cancer. Chemotherapy alone concluded to be an independent risk factor.

Exposure to a few selected industrial chemical including vinyl chloride, phenoxyacetic acid, arsenic and phenoxyherbicides may be followed by the appearance of sarcomas. There are, however, a number of inherent problems in occupational epidemiology with small number of patients in any given series and the difficulty in isolating a single agent (Sathiakumar and Delzell, 1997). For these reasons, few associations can be considered established and causal (Dich et al., 1997). For example, there is a clear association between vinyl chloride and hepatic

angiosarocoma (Lee et al., 1996). Phenoxyacetic acid (Hardell and Eriksson, 1988) and arsenic (Lander et al., 1975) are also implicated as inducing agents for hepatic sarcomas in humans. Wingren et al (1990) reported an increased incidence of soft tissue sarcomas in gardeners (phenoxy herbicides), railroad workers, construction workers exposed to impregnating agents or asbestosis and unspecified chemical workers. The association between exposure to phenoxy herbicides and soft tissue sarcoma has been corroborated (Vineis et al., 1991). The last risk may be greater 2,3,7,8-tetrachlorodibenzo-p-dioxin (TCDD) or higher chlorinated dioxins. High intensity chlorophenol exposure in jobs involving wood preservation, machinists and the use of cutting fluids may increase the risk of soft tissue sarcoma, independent of phenoxy herbicides (Wingren et al., 1990; Hoppin et al., 1998). However, some studies have not confirmed this association (Hardell and Erikson, 1988).

Chronic oedema and trauma may also be contributing factors to the malignant transformation. Sarcomas of soft tissue (primarily lymphangiosarcomas) may be observed following massive and quite protracted oedema. Classically, this has been seen in the post-mastectomy, lymphoedematous arm (Stewart-Treves syndrome) (Stewart and Treves, 1948; Tomita et al., 1988). It has also been described following chronic lymphoedema due to filarial infection (Muller et al., 1987).Chronic irritation secondary to foreign bodies may be a factor in the induction of sarcomas. Trauma is rarely a factor in the development of these tumours with the exception of desmoid tumours.

CLASSIFICATION

The rationale for developing a well-defined , comprehensive and flexible classification system of soft tissue tumours is to provide morphological guidelines which expand our understanding of neoplasia , predict biological behaviour and facilitate the development of more effective treatment. Originally, classification schemes were descriptive in nature and based on tumour cell configuration. Subsequently they have evolved through the concept of histogenesis or 'cell of origin' to the current belief that a primitive or stem-like mesenchymal cell undergoes neoplastic transformation and, depending on the genetic code translated, differentiates along one or multiple cell lines.

SOFT TISSUE SARCOMA- WHO CLASSIFICATION (MODIFIED)

1. Fibrous tumours
2. Fibrohistiocytic tumours
3. Lipomatous tumours
4. Smooth muscle tumours
5. Tumours of blood vessels and lymphatics
6. Skeletal muscle tumours
7. Perivascular tumours
8. Synovial tumours
9. Mesothelial tumours
10. Peripheral nerve sheath tumours

11. Primitive neuroectodermal tumours
12. Extraskeletal and cartilaginous tumours
13. Miscellaneous tumours

Currently, the most widely used classification system is the Enzinger and Weiss(2001) modification of the World Health Organization formulation. In this system, soft tissue tumours, including non-neoplastic tumour-like lesions, are categorized into three broad groups:

1. tumours which differentiate along cell or tissue lines that have normal counterparts, i.e., fibrous tissue, fat, vessels, smooth muscle, nerve, ganglia, synovium, bone and cartilage;
2. tumours whose lines of differentiation have no normal counterpart but which are consistent and recognized by a distinctive morphology, i.e, myxoma, epithelioid sarcoma, and alveolar soft part sarcoma; and
3. tumours which are so poorly differentiated and morphologically unique that they defy classification.

The vase majority of tumours fall into the first two groups. Overall, there are approximately 200 different entities, of which 80 are malignant.

GRADING

The histological typing of soft tissue tumours does not per se provide sufficient information on which to base therapeutic decision. Tumour grading is based on the concept that morphology reflects biological behaviour. The specific microscopic characteristic of soft tissue tumours that best predict their aggressiveness, i.e the potential for regional and distant metastasis, can be identified, integrated and represented by grade.

The 1997 American joint Committee on Cancer (AJCC) and International Union Against Cancer (UICC) staging system for sarcoma of soft tissue (Fleming et al.,1997) is based upon a four-step grading, i.e, low, intermediate, high-grade and undifferentiated neoplasms. In this grading system, grade 1 and 2 tumours are considered low grade lesions with minimal metastatic potential; grade 3-4 neoplasms are high grade tumours that carry a significant risk of metastasis. Many institution continue to employ a three-step grading scale consisting of low (G1), intermediate (G2) and high (G3) grades. In this grading scale G1 lesions are considered to have minimal metastatic potential and are analogous to the G1 and G2 designations on the four-step scale for staging purposes. The G2 and G3 tumours on the three-step scale are analogous to G3 and G4 on the four-step scale. The designation of grade is based upon a consideration and integration of each of these morphological features: degree of cellular differentiation, extent of necrosis, number of mitoses, cellularity, pleomorphism or anaplasia, quantity of matrix, vascularity, hemorrhage, vascular invasion and encapsulation . Among these variables, necrosis, mitoses and degree of differentiation appear to be the best predictors of outcome. Despite some lack of agreement on the number of grades employed and the significance of individual

morphological parameters (there is inevitably a subjective component in assigning grade and only a part of the tumour is examined) grading, more than any clinical and pathological parameter available, is the most important prognosticator.

IMMUNOHISTOCHEMISTRY

Immunohistochemistry has become increasingly important in diagnosing sarcomas (Ushigome et al.,1992). This technique is based on the utilization of visually tagged monoclonal or polyclonal antibodies directed against proteins found in specific cell types. Immunohistochemisty in soft tissue sarcoma :

ANITBODY	**DISTRIBUTION**
Vimentin	Almost all sarcomas and some carcinomas
Keratin	Almost all carcinomas and some sarcomas (Epithelioid sarcoma, synovial sarcoma)
Desmin	Leiomyosarcoma, rhabdomyosarcoma and occasionally MFH
Neurofilmanet	Primitive neuroectodermal tumour, neuroblastoma
S-100 protein	Malignant schwannoma, melanoma, clear cell (melanoma of soft parts), chondrosarcoma, leiomyosarcoma, rhabdomyosarcoma, liposarcoma
Myoglobin	Rhabdomyosarcoma

Factor VIII-related antigen	Angiosarcoma, Kaposi's sarcoma
Actin	Leiomyosarcoma, rhabdomyosarcoma (MFH)
EMA	Carcinomas, synovial sarcoma, meningioma
Leu 7	Malignant schwannoma, leiomyosarcoma, synovial sarcoma, rhabdomyosarcoma

STAGING

The Task Force on Soft Tissue Sarcomas of the American Joint Committee on Cancer (AJCC) Staging and End Result Reporting and the Union Internationale Centre le Cancer (UICC) have established a staging system for soft tissue sarcomas which is an extension of the TNM system to include G for histological grade. Grade, size, depth and presence of nodal or distant metastases are the determinants of stage. This staging system is applied to all sarcomas of soft tissue except rhabdomyosarcoma (for which there is a special staging system), Kaposi's sarcoma, dermatofibrosarcoma, desmoid, and sarcoma arising from the dura mater, brain, parenchymous organs or hollow viscera. The staging system was revised in 1997 with the addition of subgroupings of the T stage to designate superficial and deep lesion and the assignment of patients with nodal involvement to stage IV. Superficial lesions do not involve the superficial fascia in extremity lesions. For practical purposes, all retroperitoneal and visceral lesions would be deep lesions. The staging system which places primary emphasis on extension beyond compartment is useful for planning surgical approach (Enneking et

al.,1980). Compartmental status in combined modality treatment does not affect outcome to an important degree.

AJCC staging system for sarcoma of soft tissues (AJCC Staging Manual, 1997):

Histological Grade of Malignancy:	
GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated
T Primary Tumour:	
TX	Primary tumour cannot be assessed
T0	No evidence of primary tumour
T1	Tumour 5cm or less in greatest dimension T1a: superficial tumour T2b: deep tumour
T2	Tumour greater than 5cm in greatest dimension T2a: superficial tumour T2b: deep tumour
N Regional Lymph nodes:	

NX	Region lymph nodes cannot be assessed
N0	No histologically verified metastases to regional lymph nodes
N1	Histologically verified regional lymph node metastasis
M Distant metastasis	
MX	Distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis
Stage I	
A	Low-grade, small, superficial and deep: G1-2 T1a-1b N0 M0
B	Low-grade, large, superficial: G1-2 T2a N0 M0
Stage II	
A	Low-grade, large, deep; G1-2 T2b N0 M0
B	High-grade, small, superficial and deep; G3-4 T1a-1b N0 M0
C	High-grade, large, superficial; G3-4 T2a N0 M0
Stage III	High-grade, large, deep; G3-4 T2b N0 M0
Stage IV	Any metastasis; any G any T N0 M0; any G any T N0 M1

CLINICAL ASPECTS OF SPECIFIC SOFT TISSUE SARCOMAS

1. Alveolar soft parts of specific soft tissue sarcomas.
    a. Tissue of origin (incidence), Unknown (rare)
    b. Features. Unique histology with no benign counterpart; often indolent even with lung metastases, which are common. The sarcoma most associated with brain metastases. Commonly affects the thigh in adults and the head and neck in children. The 5-year survival rate exceeds 60%.

2. Angiosarcoma (hemangiosarcoma, lymphangiosarcoma)
    a. Tissue of origin (incidence). Blood or lymph vessels (2% - 3%)
    b. Feature of hemangiosarcoma. Affects the elderly; aggressive. Arises in many organs, notably the head and neck region, breast, and liver; especially affects the skin and superficial soft tissues (whereas most STSs are deep). Dedifferentiation from a hemangioma is rare. The 5-year survival rate is less than 20%
    c. Features of lymphangiosarcoma. Affects older adults; aggressive. Arises in areas with chronic lymphatic stasis (especially postmastectomy). The 5 – year survival rate is 10%.

3. Clear cell sarcoma
    a. Tissue of origin (incidence). Now recognized as a form of malignant melanoma (rare).
    b. Features. Affects adults younger than 40 years of age; painless, firm, spherical masses on tendon sheaths and aponeurotic structures of distal extremities. The 5 – year survival rate is about 50 percent.
4. Epithelioid sarcoma
    a. Tissue of origin (incidence), Unknown (rare)
    b. Features. Affects young adults; aggressive; typically appears on distal extremities. Epithelioid sarcoma and synovial sarcoma are the most common tumours of the hand and foot. Differs from other STSs by

having a greater tendency to spread to non contiguous area of skin, subcutaneous tissue, fat, draining lymph nodes, and bone. The 5- year survival rate is 30%.

5. Fibrosarcoma
    a. Tissue of origin (incidence), Fibrous tissue (5% to 20%)
    b. Features. Affects all age groups; arises in many mesenchymal sites; usually involves the abdominal wall or extremities. Ninety percent are well-differentiated (desmoid). Dermatofibrosarcoma protuberans (rare) develops on the skin of the trunk and almost never metastasizes. Fibromyxosarcoma affects any soft tissue site but usually develops on the extremities. Ten percent are poorly differentiated (high grade). Survival is directly related to tumour grade
6. Malignant fibrous histiocytoma (MFH)
    a. Tissue of origin (incidence), Unknown (10% - 23%)
    b. Features. Age older than 40 years (less than 5% of affected patients are younger than 20 years of age). MFH has become a popular histolgical diagnosis, often encompassing tumours that were previously classified as pleomorphic rhabdomyosarcoma or undifferentiated fibrosarcom; it is the most common STS in some series. MFHs range from benign to atypical to frankly malignant. Develops in extremities (especially legs), trunk, and retroperitoneum. Superficial MFH develops close to the skin surface and is often low grade; the 5 – year survival rate is

65%. Deep MFH usually is high grade; the 5- year survival rate is 30% to 80%.

7. Hemangiopericytoma/solitary fibrous tumour
    a. Tissue of origin (incidence), Blood vessels or fibrous tissue (less than%).
    b. Features. Affects all ages. Develops under finger tips (glomus tumours), on lower extremities or pelvis, in the retroperitoneum, and elsewhere. Benign and malignant versions. The 5 – year survival rate is about 50%.

10. Liposarcoma

    c. Tissue of origin (incidence), Fat tissue (15% - 18%)
    d. Features. Affects middle and older age groups, mostly men. Develops in thigh, groin, buttocks, shoulder girdle, and retroperitoneum. Does not arise from benign lipomas. The 5 – year survival rate is 80% for low grade liposarcomas and 20 % for high-grade liposarcomas.

11. Mesothelioma
    a. Tissue of origin (incidence). mesothelium
    b. Features. Age older than 50 years. Asbestos exposure is etiologic. Involves pleura and peritoneum; aggressively encases viscera. Highly lethal; the 5 – year survival rate is less than 10%.
12. Myxoma
    a. Tissue of origin (incidence). Mesenchymal tissues

b. Features. Usually found on extremities; has histological appearance of umbilical cord. The 5 – year survival rate is about 80%.

13. Neurofibrosarcoma (schwannoma, neurilemoma)

a. Tissue of origin (incidence). Nerve(5% - 7%).

b. Features. Affects young and middle-aged adults and patients with neurofibromatosis type 1 (von Recklinghausen's disease; about 10% develop sarcomatous changes during lifetime). Histologically resembles fibrosarcoma. Presents with thickening of nerves and without anatomic predilection. Superficial variety is low grade, spreads extensively along nerve sheaths without metastasizing, and has a 5–year survival rate of more than 90%. Penetrating variety is high grade with nodular growth, vascular invasion, and lung metastases and has 5-year survival rate of less than 20%

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14. Rhabdomyosarcoma

a. Tissue of origin (incidence). Straited muscle (5%-19%).

b. Features. By definition in the G-TNM staging system, all are grade3. All types can occur in any age group.

c. Features of embryonal rhabdomyosarcoma. Affects infants and children; sites are head and neck (70%) and genitalia (15%-20%). Includes sarcoma botryoid. The 5-year survival rate is about 70%.

d. Features of alveolar rhabdomyosarcoma. Affects teenagers at any site; highly aggressive; histology resembles lung alveoli. The 5-year survival rate is about 50%.

e. Features of pleomorphic rhabdomyosarcoma. Affects patients older than 30 years of age, is rare, and develops in extremities. Often is highly anaplastic; microscopically confused with MFH. The 5-year survival rate is about 25%

15. Synovial sarcoma

a. Tissue of origin (incidence). Tenosynovial mesothelium(5%-20%)

b. Features. Affects young adults, but may occur from the second to fourth decade. Monophasic and biphasic subtypes are distinguished. Presents with hard masses, often painful, near tendons in the vicinity of joints of the hands, knees, or feet. Synovial and epithelioid sarcoma are the most common tumours of the hand and foot, often calcified, with characteristic radiographic appearance. The majority of synovial sarcomas are high grade. Lymph node involvement may be seen in up to 20% of cases. The 5-year survival rate is from 30%-50%.

CLINICAL EVALUATION

CLINCAL HISTORY

The most frequent initial complaint is that of painless lump of a few weeks to several months duration. Occasionally, pain or tenderness precedes the detection of a lump. With progressive growth of tumour, symptoms appear which are secondary to

infiltration of or pressure on adjacent structures (e.g, tendons, muscles, nerves) or organs. Occasionally symptoms secondary to the metabolic effects of the tumour products are seen, e.g, fever, anaemia, lethargy, weight loss, histamine-like reactions. These are not rare in patients with malignant fibrous histiocytoma (Enzinger and Weiss, 2001). To get clinical genetic data in sarcoma patients, the history should include details of the cause of death and history of malignant disease in siblings, parents, grandparents and progeny.

ANATOMICAL SITE, SEX AND AGE

Sites of appearance of soft tissue sarcoma in order of frequency are; lower extremity, upper extremity, torso, head/neck, retroperitoneum. There is only a very slight preponderance of soft tissue sarcoma in males. They are more common in older people, with 40 percent in persons > 55 years of age and 15 percent in patients < 15 years of age (Enzinger and Weiss, 2001), Rhabdomyosarcomas almost always arise in children, synovial sarcomas develop in late adolescence and young adulthood and liposarcoma and malignant fibrous histiocytoma usually occur during mid and late adulthood.

PHYSICAL EXAMINATION

There must be a complete physical examination with particular attention paid to the region of the primary lesion : definition of size, site of origin (superficial or deep, attached to or fixed to deep structures), solitary or multinodular, involvement or discoloration of overlying skin, functional status of vessels and nerves, presence of

distal oedema, muscular strength, range of motion of affected part, etc. If the patient has had prior excision, the operated site should be examined for presence of ecchymosis, status of wound healing, palpable evidence of residual tumour and location of drain site. The regional and distant lymph-node groups need to be examined with care in all patients, especially those with large grade 2 and 3 (on a three step grading scale) sarcomas. Involvement of regional nodes is relatively frequent in patients with rhabdomyosarcoma and epithelial sarcoma, but uncommon in patients with fibrosarcoma and malignant fibrous histiocytoma. Myxoid liposarcoma not uncommonly metastasizes first to soft tissue or bone.

NATURAL HISTORY

LOCAL RECURRENCE

One of the major clinical problems when treating soft tissue sarcomas is the propensity of the primary tumour to recur locally. Soft tissue sarcomas enlarge in a centrifugal fashion and compress normal tissue, giving the appearance of encapsulation. This pseudocapsule is actually composed of an inner compressed rim of normal tissue (compression zone) and an outer rim of edema and small newly formed vessels (reactive zone). Fingers of tumour can extend into and through this pseudocapsule provides surgeons with a tempting plane for dissection and invites a shell out procedure, such an excision leaves microscopic and often gross tumour in the wound. Excision of any sarcoma in the pseudocapsule is inadequate therapy and results in the development of local recurrences in up to 90% of patients.

The site of a soft tissue sarcoma can certainly influence the technical ease with which resectability can be accomplished, hence it affects the potential for local control. For instance; lesions of the head and neck regions, where abutment to vital structures is often the case, are less likely to be controlled than lesion in the extremities. In the extremity, the site of the tumour may also have prognostic implications. Local control may be more difficult to achieve for proximal tumours than those more distally located, possibly because they achieve a larger size before detection. Simon and Enneking reported local recurrences following surgery alone in the buttock, groin, thigh, and areas below the knee to be 38%, 14%,15% and 0%, respectively. Potter et al, reported that among 115 patients with thigh sarcomas disease-free and overall survival was significantly worse for patients with tumour spread into the upper half of the thigh than for those with involvement limited to the distal thigh. In contrast, Yang et al found that patients undergoing multimodality treatment for sarcomas in the popliteal, axillary , or antecubital fossa had local control and overall survival rates similar to those for patients with purely intracompartmental extremity tumours. Hence the significance of location in the extremity may be less with multimodality therapy than with surgery alone.

The time to local recurrence following surgery follows a predictable pattern. In one series of patients treated with surgery alone, approximately 80% of all lesions that were destined to recur locally did so within 2 years.

Isolated local recurrences of soft tissue sarcomas, regardless of site, should be considered for further resection. A thorough evaluation for the presence of

disseminated disease should be performed before contemplating resection of a local recurrence.

DISTANT METASTASIS

Despite adequate local control of the primary lesion, many patients with high grade and a few patients with low grade soft tissue sarcomas succumb to metastatic disease. Although most patients with soft tissue sarcomas present without obvious metastases, many harbor occult micrometastasis that eventually become clinically evident. These patient represent a population who could benefit from adjuvant systemic chemotherapy along with removal of the primary tumour.

As with local recurrence, the incidence and pattern of distant metastasis depends on the site of the primary sarcoma. Patients with retroperitoneal sarcomas had a greater tendency for local recurrence and disseminated disease throughout the abdomen. Patients with head and neck and truncal sarcomas had a higher local recurrence rate than those with extremity sarcomas. Treatment options for patients with metastatic disease include surgical resection if the tumour is localized to one organ system or systemic cytotoxic chemotherapy if it is not.

LABORATORY INVESTIGATIONS

Laboratory studies need not go beyond a complete blood count and urinalysis for all but the exceptional patient. For patients with lesions in the abdomino-pelvic cavity,

liver function and/or liver scan should be obtained as involvement of liver is not rare in such patients.

IMAGING MODALITIES EMPLOYED TO EVALUATE SOFT TISSUE TUMOURS

CONVENTIONAL RADIOGRAPHIC STUDIES

The plain film radiograph obtained with low kilovoltage technique (below 50kvp) enhances the difference in radiographic density between fat and muscle and accentuated the soft tissue detail. Routine or over penetrated radiographs best demonstrate involvement of adjacent bone. Plain tomography is of particular value for assessing calcifications in soft tissue masses and for delineating soft tissue abnormalities adjacent to complex bony structures (pelvis, spine, chest wall, shoulder). Plain tomography augments but does not replace the plain radiograph The plain film is still invaluable for predicting the presence and nature of bony involvement. When adjacent bone is involved by a slowly growing soft tissue mass, local pressure by the mass results in a scalloped pattern with a well define sclerotic margin which is most frequently encountered in benign process. Irregular cortical destruction is usually associated with fast-growing, frequently malignant lesions. Obliteration or displacement of normal fascial planes due to soft tissue infiltration is common in both benign and malignant tumours. Occasionally this interface is lobulated and smooth, suggesting the presence of a confining capsule. Few soft tissue tumours have a true capsule, and what resembles a capsule is imply a compressed rim

of normal structures. Thus a smooth margin can be deceptive if it is automatically equated with benignancy.

SONOGRAPHIC STUDIES

Sonography provides an accurate method for detecting and determining the size of the soft tissue mass in the extremities due to differential patterns of echoes detected in the mass compared to those in normal muscle and fascia. This modality is far less reliable than CT or MR when the mass occurs in complex anatomic locations such as the pelvis, is located in deep soft tissues adjacent to bone, or occurs in the thorax surrounded by the lung. When the soft tissue mass occurs in other locations, particularly in the abdomen or extremities, sonography precisely establishes anatomic relations and tumour margins but generally provides considerably lesser anatomic detail than in available CT or MR. Most soft tissue tumours have an echogenicity distinct from the adjacent soft tissues, which provides an interface between the tumour and surrounding soft tissues. The size and configuration of the mass can be accurately determined by sonography because the lesion can be visualized in both the transverse (axial) and longitudinal (sagittal) planes. Therefore sonography may be useful for following the size of the soft tissue mass on serial studies or the response of the lesion to treatment by non surgical means such as chemotherapy or radiation therapy. The echo pattern of solid masses is nonspecific, but a fluid-filled mass such as a synovial cyst (popliteal cyst) can be easily differentiated from a solid mass by sonography. Perhaps the best utility for sonography is an ultrasound-guided biopsy of a relatively superficial soft tissue mass.

ANGIOGRAPHIC STUDIES

Angiography is most helpful for evaluating the vascular supply of a soft tissue mass and the impact of the mass on adjacent structures. The angiogram typically performed with the digital subtraction technique (digital subtraction angiography, or DSA), provides information about the anatomic extent of the tumour, including its effect on adjacent structures, the arterial supply and venous drainage. Because these features influence the choice of operative procedures, angiography is a valuable adjunct to patient management. Much of this anatomic information can also be obtained in a noninvasive fashion via magnetic resonance angiography (MRA). Because the patient with a soft tissue tumour is likely to undergo MR scanning anyway, the referring physician should discuss with the radiologist the appropriateness of performing MRA as a supplement to the routine MR pulse sequences. Interventional angiographic procedures are sometimes necessary, occasionally to embolize a highly hypervasuclar soft tissue mass preoperatively or, rarely, to embolize a persistent arterial bleeder following surgical resection of the lesion.

Malignant soft tissue tumours have angiographic patterns that range from hypervascular to hypovascular. The angiographic features of neovascularity, puddling, and tumour blush are typical of malignant tumours, although benign tumours also occasionally demonstrate neovascularity. Malignant soft tissue tumours are frequently heterogeneous and contain less-vascular areas, corresponding to areas of hemorrhage and necrosis along with hypervascular areas that correspond to viable

tumour. Unsuspected satellite tumours adjacent to a primary mass may also be discovered by angiography.

Certain angiographic patterns are suggestive of specific histologic diagnoses such as hemangiomas and vascular malformations. Hemangiopericytoma is another tumour that may exhibit a unique and striking hypervascular pattern. A frequent angiographic feature of hemangiopericytoma is that early during the arterial phase the main arteries are displaced around the periphery of the tumour. Later, the feeding arteries spread in a meshwork pattern before penetrating the lesion. This peripheral vascular distribution noted at angiography correlates well with the plexiform meshwork of vessels covering the tumour seen on inspection of the gross specimen. This angiographic pattern is highly suggestive of hemangiopericytoma. In most soft tissue masses, however, a specific pathologic diagnosis cannot be predicted reliably by angiography.

CT SCANNING

Computed tomography has proved helpful for detecting soft tissue tumours and monitoring the patient for local recurrence and distant metastates. Therefore if a patient is to undergo CT scanning or MR imaging, it is imperative that it be done prior to any biopsy or surgical intervention. If this principle is ignored, the value of the combined imaging studies is dramatically diminished because it becomes difficult, or even impossible to assess how much of the soft tissue abnormality is due to the lesion versus surgically induced changes such as edema and hemorrhage. CT scanning is most useful for defining the extent of the soft tissue mass and its relation

to adjacent structures, especially in complex anatomic sites such as the pelvis. This exquisite anatomic detail enables the surgeon to perform more complete resection of the tumour and the radiotherapist to define a more precise radiation therapy field. Occasionally, CT scanning can help establish a diagnosis, especially with regard to detection of adipose tissue in tumours whose fatty component may not be appreciated on plain radiographs. Contrast enhancement is valuable for assessing the vascularity of the soft tissue mass and distinguishing the mass from adjacent structures, especially the vascular bundle. Soft tissue tumours can also be isodense with normal muscle on precontrast CT scans and become evident only following contrast infusion. CT scanning is superior to MR imaging for detecting and characterizing calcification in the soft tissue mass and detecting subtle cortical erosion and periosteal reaction in the bone adjacent to the mass. The ability of CT scanning to detect calcification in the soft tissue mass can prove invaluable for suggesting a specific diagnosis. For example, phleboliths suggest hemangiomas; calcified rings and arcs suggest a cartilaginous lesion; amorphous calcification in a large soft tissue mass suggests a soft tissue osterosarcoma or synovial sarcoma; and peripheral calcification surrounding a soft tissue mass suggests myositis ossificans, whereas calcification throughout the lesion favours tumoral calcinosis.

MAGNETIC RESONANCE IMAGING

Magnetic resonance imaging is arguably the most important technologic innovation in medicine during the last 15 years. Both the CT and MR are expensive and depend on sophisticated computers that rapidly analyze millions of complex differential

equations. There are significant differences between the two methods. A major advantage of MR imaging is its ability to image the region of anatomic interest directly in orthogonal planes (i.e., axial, coronal, and sagittal planes). This capability and the excellent soft tissue contrast of the image are invaluable when assessing the extent of a soft tissue mass.

Most soft tissue tumours exhibit a low signal intensity (i.e., they appear dark) or intermediate signal intensity on T1-weighted scans; and they exhibit a high signal intensity (i.e., they are bright or white) on T2 weighted scans. Therefore MR provides little diagnostic specificity for most soft tissue tumours, as explained earlier in the chapter. However, one group of tumours typically have a high signal on T1-weighted scans, including hematomas, lipomas, liposarcomas, hemangiomas, and lesions complicated by hemorrhage into a preexisting tumour. The histologic picture of these lesions varies, but all contain blood or fat. In contrast, flowing blood, unlike the stagnant blood in a hematoma, has a low signal intensity, permitting assessment of vessel patency. Lesions that are predominantly fibrous exhibit low signal intensity on both T1- and T2-weighted images.

Most soft tissue masses have a nonspecific appearance on MR imaging; but even so, this technique is the imaging modality of choice for determining (in three orthogonal planes) the extent of the lesion. If the plain radiograph obtained from the patient with a palpable soft tissue mass fails to demonstrate bony involvement or calcification in the soft tissue lesion, only MR imaging (not CT scanning) need be performed.

BIOPSY

Properly performed, a timely biopsy is the critical first step in multimodality treatment approach. Improperly done, it can complicate patient care and sometimes even eliminate treatment options. Several biopsy techniques are available to the clinician: fine needle aspiration, core-needle biopsy, incisional biopsy, and excisional biopsy. The choice of biopsy is dictated by the size and location of the mass and the experience of the pathologist. Excisional biopsy should be reserved for small (<3-5cm in greatest diameter) and superficial soft tissue masses where the chance of malignancy is low and where complete excision would not jeopardize subsequent treatment in the event a sarcoma is found.

Fine-needle aspiration (FNA) involves use of a fine-gauge needle (usually 21-23 gauge) to aspirate individual tumour cells from a mass. FNA cytology has a role to play in the diagnosis of some soft tissue lesions, but its use should be limited because even experienced cytopathologists are often unable to discern the grade and histologic type of a sarcoma from the small cellular sample of an aspirate. The advantage of FNA is that it is relatively atraumatic and hence can be used to sample deep-seated tumours (e.g., retroperitoneal masses) under computed tomography (CT) guidance. FNA minimizes the potential for tumour spillage in the peritoneal cavity that can accompany open surgical biopsy of a retroperitoneal sarcoma. CT-guided FNA has proven helpful for diagnosing intraabdominal and retroperitoneal tumours but is rarely needed for extremity sarcomas. FNA biopsy is also acceptable for documenting local or distant recurrence in patients with a previously diagnosed sarcoma, where the cytology findings can be directly compared with the prior histology specimens.

Core-needle biopsy results in the retrieval of a thin sliver of tissue (app. 1×10m). Here again, the small sample size may make it difficult for a pathologist to diagnose the tumour accurately, or the tissue obtained may not be representative of the entire tumour, leading to an underestimate of the grade. Tissue necessary for special stains or electron microscopy may not be available with this technique. Previously expressed fears that core-needle biopsy of extremity sarcomas would result in a significant number of hematomas – and hence the dissemination of tumour cells beyond the confines of the primary lesion – appear groundless. Several series have compared core-needle and open biopsies of soft tissue tumours and documented that both the histologic type and grade of a sarcoma could be correctly determined by core-needle biopsy in about 90% of cases. Moreover, the core-needle biopsy approach is more cost-effective. These results have encouraged wider use of this technique, including core-needle biopsies using CT guidance.

Excisional biopsy refers to removal of the entire grossly evident lesion, usually without a significant margin of normal tissue. Many sarcomas appear to be encapsulated at the time of open biopsy. In actuality, these tumours have a "pseudocapsule", and removing the tumour through its pseudocapsule leaves gross or microscopic cancer behind in many cases. "Shell out," or excisional biopsy, should be reserved for lesions less than 3-5cm in diameter or for extremely superficial tumours. Excisional biopsies of large or deep sarcomas are undesirable as they can contaminate surrounding tissue planes, which may compromise the subsequent definitive surgical procedure.

Incisional biopsy involved removal of a generous wedge of tissue that is minimally manipulated at the time of surgery. There are several important technical factors when performing an incisional biopsy. For extremity lesions, the incision is oriented along the long axis of the extremity. For truncal or retroperitoneal lesions, the biopsy incision is situated so it can be readily excised along with the tumour if a sarcoma is diagnosed. The biopsy site should be directly over the tumour, at the point where the lesion is closest to the surface, and there should be no raising of flaps or disturbance of tissue planes superficial to the tumour. Prior to wound closure, careful attention is paid to hemostasis to minimize the likelihood of a hematoma, which could disseminate tumour cells through normal tissue planes. Drains are not used routinely; in the uncommon case where a drain is required, it should exit either though or near the biopsy incision. If malignancy is diagnosed, the drain tract must be excised in continuity with the tumour mass.

A frozen section examination at the time of biopsy can be useful for determining whether the specimen contains viable lesional tissue for examination, in contrast to nonviable tumour or nonlesional reactive tissue surround the tumour. Because of the subtle criteria necessary to distinguish benign from malignant soft tissue tumours and the importance of proper grade assignment, treatment planning (including definitive resection) is almost always deferred until a definitive report based on permanent sections is available.

TREATMENT OF LOCALIZED TUMOURS

SURGICAL THERAPY

Surgery is the mainstay of any treatment approach for a clinically localized primary or recurrent soft tissue sarcoma. As has been described, however, removal of the gross tumour mass from within its pseudocapsule is associated with a prohibitively high likelihood of local recurrence. With the appreciation of the infiltrative nature of these tumours, radical procedures such as amputation came to be accepted as the standard for surgical therapy. Enneking classified surgical procedures for extremity sarcomas according to the margins achieved. He described four types of excision.

1. Intracapsular excisions are performed inside the pseudocapsule and are often piecemeal in nature. An amputation that passes within the pseudocapsule is called an intracapuslar amputation. The likelihood of local recurrence with intracapsular procedures is virtually 100%.
2. Marginal excisions are en bloc resections performed through the tumour. Excisional biopsies and “shellout” procedures fall into this category. An amputation performed though this marginal zone is called a marginal amputation. Local recurrences are expected 60-80% of the time.
3. Wide excisions are an en bloc resections done through normal tissue beyond the reactive zone but within the muscular compartment of origin, leaving place some portion of that compartment. The pseudocapsule is removed en bloc, and the tumour is never visualized during the procedure. The reported local recurrence rate following wide excision alone varies depending on selection criteria and the adequacy of the margin as assesed histologically, but overall it is approximately 30%. An amputation can be considered a wide

amputation if it is performed though normal tissue proximal to the reactive zone but remains within the compartment of involvement.

4. Radical excisions are en bloc resections of the tumour and the entire compartment of origin, leaving no remanents of the compartment intact. A radical amputation usually requires disarticulation of the joint proximal to the involved compartment and results in removal of the entire compartment at risk.

COMPARTMENT EXCISIONS

For tumours that arise entirely within an anatomically defined muscular compartment, excision of the muscles comprising that compartment from origin to insertion is often successful in achieving local control. The thigh, which is the site of more than 50% of all extremity sarcomas, has three major compartments bound by the fascial at and its extension. The anterior compartment includes primarily the quadriceps and sartorius muscles, which can be removed along with the femoral nerve. An anterior compartment excision results in knee weakness and instability, which can be improved with the use of the transplanted gracilis and short head of the biceps. Even without such reconstruction, patients can ambulate after an anterior compartment excision by using the hip flexor to throw the leg forward; gait is improved with the use of a locking knee brace. The medial thigh compartment consists of the gracilis, adductors (minimus, brevis, longus, and magnus), and pectineus muscles. Medial compartment excision is generally well tolerated with only limited functional deficit.

Posterior compartment excision removes the hamstring muscles (semimembranous and semitendinosus and biceps femores), as well as the posterior portion of the adductor magnus muscle. Ambulation is surprisingly well maintained after this surgery, with knee flexion brought about by the action of the gracilis and soleus muscles on the distal femur. Buttockectomy entails removal of the entire gluteus maximus muscle for tumours localized entirely within this muscle.

LIMB-SPARING PROCEDURES

Rosenberg et al, compared radical amputation to wide local excision plays postoperative irradiation in a prospective, randomized trial. Patients who underwent limb-sparing surgery had a survival rate identical to those undergoing amputation, despite a slightly higher local recurrence rate (19% Vs 6%, a difference that was not statistically significant). This study demonstrated the merit of lib-sparing approached to extremity sarcomas. Radical amputation is currently reserved for patients who are not suitable candidates for limb-sparing approaches, usually because of abutment to bone or major neurovascular structures or an extremely large tumour size. It is of note that the location of the tumour distally in the extremity (hand or foot) is not necessarily an indication for amputation. Local control with acceptable with limb-sparing approaches in most of these cases.

Most current limb-salvage protocols include wide excision as the definitive surgical procedure. Wide excision involves gross total removal of the tumour with a wide margin of normal tissue, but no attempt is made to resect an entire muscle compartment. Rather, a margin 3-5cm of normal tissue is obtained proximally and

distally. It includes excision of some overlying skin to encompass all previous scars or biopsy sites. Tumour should not be visualized during the excision so tumour cells are not spilled into the surgical bed. On the lateral deep margins, at least one grossly uninvolved fascial plane is resected en bloc with the tumour for large or deep-seated tumours, resection of uninvolved periosteum or adventitia my represent the deep margin. Removal of periosteum should be limited to areas directly abutting tumour to minimize the likelihood of post irradiation pathologic fracture. If necessary, major vascular structures may be resected and reconstructed with graft material. On occasion, major nerves (e.g., sciatic nerve) are sacrificed to preserve a functional albeit neurologically compromised extremity. Because of their less aggressive nature, when low grade lesions are resected major vessels or nerves are not taken along with the tumour. Placement of titanium clips outlining the limits of the excision is essential as a guide to the radiation therapist for constructing the radiation treatment portal. Suction catheters are placed at the end of the dissection to allow evacuation of any blood and serous fluid from the operative bed and to promote the adherence of skin flaps.

The use of multimodality therapy (particularly preoperatively) is associated with a high incidence of wound complications. These complications can be disastrous in an irradiated wound or if major vessels, nerves or bone become exposed. To minimize the likelihood of wound breakdown, consideration should always be given to reconstructing the surgical defect with free or pedicled myocutaneous flaps. Barwick et al, articulated the criteria for primary closure of a wide excision wound in patients undergoing multimodality therapy. The skin edges should be approximated without

tension, and the resection site must not have exposed bone, nerve, blood vessel, or tendon present. If the skin edges cannot be approximately without tension but the base of the resection is entirely muscle, a split-thickness skin graft should be applied; otherwise, a flap reconstruction is performed. Irradiated wounds in the trunk and retroperitoneum may present a formidable technical challenge to the reconstructive surgeon.

RADICAL AMPUTATIONS

Below-knee amputation is performed through the tibia and fibula and allows a stump to be used for fitting a prosthetic device. Rehabilitation from this procedure is usually satisfactory. Below-knee amputation is the radical resection for any soft tissue sarcoma of the foot. Above-knee amputation can be performed at any level distal to the lesser trochanter to allow enough stump for a prosthesis. This amputation does not constitute a radical excision for tumours above the knee, as the entire compartment is not removed. Hip disarticulation entails complete removal of the femur at the hip joint. Most muscles attached to the lower extremity are removed in their entirety. This procedure constitutes one option for radical excision for patients with lesions of the middle and lower thigh. Hemipelvectomy involves removal of the entire lower extremity and hemipelvis removal of the entire lower extremity and hemipelvis, with disarticulation of the sacroiliac joint and pubic symphysis. This procedure is a radical amputation for patients with proximal thigh and buttock tumors. The standard hemipelvectomy utilizes a posterior flap of skin and subcutaneous tissue overlying

the buttock. For buttock and posterior thigh lesions, it is possible to construct an anterior myocutaneous flap based on the quadriceps muscles and superficial femoral artery to cover the surgical defect. Modified hemipelvectomy preserves the iliac wing, which allows improved patient rehabilitation. It is similar to the standard hemipelvectomy except the sacroiliac joint is preserved and the iliac bone is divided below the level of the sciatic notch. Because this involves transaction of muscles in the buttock, it is not suitable for lesions in this area. Internal hemipelvectomy is not generally employed for soft tissue tumours; it involves removal of the hemipelvis without amputation of the extremity and can be useful for management of bony tumours of the hemipelvis.

For tumours of the hand and wrist requiring radical amputation, below-elbow amputation is used. Above-elbow amputation is used for tumours of the forearm. Shoulder disarticulation is reserved for distal arm and elbow lesions. Forequarter amputation (interscapulothoracic amputation) is applied to the treatment of lesions of the axilla, shoulder girdle, or proximal arm. This procedure includes removal of the entire upper extremity along with the scapula and clavicle.

The means required to achieve a radical excision depend in part on the anatomic location of the tumour and its size. Many tumours of the extremities are not located in distinct anatomic compartments, and hence a radical excision cannot be achieved except by amputation. Similarly, large tumours localized to one compartment but that abut bone or major neurovascular structures cannot be radically excised without amputation. A radical excision, as defined by Enneking, Is not required for all

sarcomas. With the use of adjuvant radiation therapy, local control rates with less radical resections have improved and match those achievable with radical surgery. Currently, wide excision is the procedure of choice for low grade sarcomas or higher grade tumours that are to be treated with multimodality therapy. Marginal excision is frequently all that can be achieved for treatment of retroperitoneal and head and neck sarcomas; intracapsular excision is not generally performed except for the occasional low grade tumour for which any other type of excision would cause an unacceptable loss of function. Adjuvant irradiation should almost always be considered when tumours must be treated with marginal or intracapsular excisions.

Overall local control was obtained in approximately 80% of patients undergoing radical resection regardless of whether amputation was performed. Unfortunately, even a "radical" excision is not always associated with an adequate margin when the resected specimen is examined histologically. When histologic evaluation verified that adequate margins were obtained with the radical procedure, the local failure rate was 5%. If microscopic tumour was found at or within 1 mm of the surgical margin, the local failure rate rose to 89%. Inadequate margins were most commonly associated with large proximal thigh and groin tumours. More distal extremity tumours rarely had tumour at or close to the surgical margin after appropriate radical excision and the local control rate can be expected to approach 100% after radical amputation of distal extremity sarcomas.

Although radical excisions that achieve a histologically negative margin are associated with high rates of local tumour control, the functional, psychological, and

cosmetic costs can be high. For this reason, radical surgery as single modality therapy has gradually been replaced by more conservative resection performed as part of multidisciplinary, multimodality treatment approaches. Nowhere is this trend more evident than during treatment of extremity soft tissue sarcomas with multimodality, limb-sparing therapy.

RADIATION THERAPY

POSTOPERATIVE RADIATION THERAPY

Postoperative radiation therapy after wide excision provides excellent local control for primary extremity sarcomas up to 10 cm in size. The randomized trial of amputation versus wide local excision previously cited validated the concept of limb-sparing surgery combined with postoperative irradiation. Generally, a total of 60 Gy or more is required to ensure local control. At these dose levels the entire circumference of the extremity must not be irradiated, in order to avoid massive lymphedema. In practice, a strip of skin and subcutaneous tissue away from the tumour is excluded from the treatment field to prevent this complication.

Radiation can also be delivered to the tumour bed postoperatively by means of implanted radioactive sources, a technique referred to as brachytherapy. This approach has the advantage of a much shorter time to initiation and completion of therapy (usually begun within a week of operation and completed in 4-5 days, compared with 6-7 weeks of external beam radiation beginning a month or more postoperatively). On the other hand, brachytherapy is technically complex and requires the presence of an experienced radiation oncologist in the operating room. A randomized trial demonstrated a significant decrease in local recurrences for high grade sarcomas after combined surgery and postoperative brachytherapy compared with surgery alone. Patients with low grade sarcomas did not benefit from adjuvant brachytherapy, although a recent effectively decreased local recurrence of these tumours. Otherwise, brachytherapy and external beam radiation appear to be equivalent when properly administered. The date from these two randomized trials provide strong support for the routine inclusion of radiation therapy (by some technique) in all patients with high grade, and many if not most patients with low grade, extremity sarcomas undergoing limb-sparingly surgery.

PREOPERATIVE RADIATION THERAPY

Preoperative irradiation followed by conservative surgery offers several theoretic advantages. The treatment volume is restricted to the known or probable extension of tumour. This means that a smaller volume can be treated than is the case with postoperative irradiation, as the latter must cover all tissues manipulated or handled during the surgical procedure. The resection may be of a lesser magnitude if tumour

regression is obtained with preoperative irradiation. Seeding the surgical bed with viable tumour cells may be reduced. Tumours initially considered unresectable without amputation, shrunk sufficiently to permit limb-sparing resection.

Enneking and McAuliffe have reported their experience utilizing preoperative irradiation followed by wide or marginal resection of low and high grade extremity sarcomas. They compared their results with comparably staged, matched control who underwent either a limb-sparing procedure or amputation without preoperative irradiation. Local recurrence rates were highest in the group undergoing limb-sparing surgery alone (37%) and were significantly higher than in those undergoing preoperative irradiation plus surgery (3%). In the same series, patients undergoing limb-sparing procedures reported substantially better postoperative function than those with amputation. The use of preoperative treatment for large extremity sarcomas has increased the number of patients who can be considered for limb-sparing surgery.

ISOLATED LIMB PERFUSION

Hyperthermic isolated limb perfusion with chemotherapeutic agents has been tried as another way to control very large tumours that would otherwise require amputation because of proximity to nerve or blood vessels. Most such regimens do not appear superior to surgery plus radiotherapy. However, hypothermic TNF-α plus melphalan has achieved complete responses rates of approximately 30%,partial remission rates of 50 % with overall limb-salvage rates over 80% (Schraffordt Koops et al., 1998).

This work was initiated by Lejeune et al, (1989) and has been extended to several other centers. TNF-α has long been known to be potent non-specific tumour cell cytotoxic agent with demonstrated high efficacy in treatment of tumour-bearing rodents. Application in man has been severely restricted due to systemic toxicity. However, by the use of the isolated limb perfusion technique, the systemic toxicity has been effectively bypassed. Effermont et al (1996) reported the result from a multi-centre study of isolated limb perfusion with TNF-α, interferon-γ (IFN γ) and melphalan for extremity sarcomas. The complete response (CR) and partial response (PR) rates were 18% and 64%; limb salvage was achieved in 84% of patients. Of 39 patients who also had conservative resection, there have been five local failures. Vaglini et al, (1994) reported from Milan a high response rate using the same protocol.

ROLE OF ADJUVANT CHEMOTHERAPY

Although surgery and radiotherapy achieve control of the primary tumour and cure most adult patients with soft tissue sarcomas, many patients, especially those with large grade 2 or 3 primaries, die of metastatic disease not evident at diagnosis. Several studies have shown chemotherapy to be effective against clinically evident metastases .This has suggested to many that chemotherapy used as an adjuvant therapy (in high-risk stage MO patients) would inactivate micrometastases and increase long-term survival.

The first prospective randomized trial of adjuvant chemotherapy for this group of tumours was reported form the M.D.Anderson Hospital (Lindberg et al.,1977); they

failed to demonstrate any advantage of vincristine, cyclophosphamide, Adriamycin and dactinomycin chemotherapy over untreated controls. Similarly, a randomized Mayo Clinic series (Edmonson et al., 1984) found a slight extension of disease-free survival with no increase in overall survival for patients receiving vincristine, cyclopohosphamide and dactinomycin alternating with vincristine, doxorubicin and dacarbazine (DTIC).

However, using Adriamycin (530mg/m^2) and cyclophosphamide (500-700 mg/m^2 Followed by high-dose methotrexate, Rosenberg et al, (1983) demonstrated a significantly improved disease-free survival for patients with extremity lesions randomized to adjuvant chemotherapy (92% Vs 60% at e years with a survival advantage of 95% Vs 74%). Some serious cardiotoxicity was encountered. With longer follow-up in this small randomized trial, the survival advantage was no longer significant (Chang et al., 1988). Single-agent doxorubicin has failed to show an improvement in disease-free or overall survival in patients receiving postoperative chemotherapy compared with surgery alone.

A metastasis of 13 randomized trials of adjuvant chemotherapy versus control in soft tissue sarcomas demonstrated that Adriamycin-based chemotherapy yielded an absolute gain in overall recurrence-free survival of 10% from 45% to 55% (P= 0.0001) and a trend for improvement of overall survival of 4% from 50-54% (P=0.12). For local control the gain was 6% at 10 years, viz. 75% -> 81%, P = 0.016. the most clear evidence of a gain in survival obtained for patients 31-60 years old, recurrent lesions, extremity and high grade (Sarcoma Meta-analysis Collaboration,

1997). Encouraging results with more intense adjuvant therapy was reported by investigators at Massachusetts General Hospital who evaluated doxorubicin, ifosfamide and DTIC chemotherapy and radiation in adult patients with > 8cm AJCC (1992) stage IIB or IIIB soft tissue sarcomas of the extremity (Spiro et al., 1996; DeLaney et al., 201). Moreover, a newer randomized trial of more intense adjuvant chemotherapy with epirubicin, ifosfamide, mesna and granulocyte colony stimulating factor (G-CSF) also seems to show a possible further advantage in disease-free and overall survival (Frustaci et al., 2001).

DISEASE (METASTATIC STAGE IVB)

CHEMOTHERAPY

Doxorubicin and ifosfamide have been demonstrated to be the most active chemotherapy agent in widely disseminated soft tissue sarcoma. For doxorubicin, objective response rates between 20% and 40% for the single agent have been reported. A steep dose-response curve for objective responses was described by O'Bryan et al, (1977)' 0% at 45mg/m^2 , 20% at 60mg/m^2 and 37% at 75mg/m^2. Complete responses were few, durations averaged 8 months with little survival advantage conferred by single-agent treatment. DTIC by itself has a modest response rate around 16% (Gottlieb et al., 1976). Methotrexate was described as active in 36% of 41 patients with six complete responders (Subramanian and Wiltshaw, 1978). High-dose methotrexate may be active as initial therapy but of limited value in pre-treated patients (Karakousis et al., 1980). Cyclophosphamide appears less active in adults than in children and less active than the related compound ifosamide (Branwell

et al., 1993). High dose of ifosfamide may be particularly active (Christmas et al., 1993”),, Cisplatin may have a role (Karakousis et al., 1979; Grabois et al., 1994) Pinede and Verweij (1986) and Greenall et al,(1986) have reviewed numerous other agents evaluated in small trials.

Many combination chemotherapy regimens for metastatic disease have been studied in phase II trials. Most of these trials include doxorubicin (or epirubicin) and an alkylating agent. Adding DTIC to doxorubicin improved the response rate to 41% as described by Gottlieh et al, (1972), but the response rate has decreased over time (Gottlieh et al, 1976). Randomized trials (Omura et al., 1983; Borden et al., 1987) found some gain f\or the combination. A South-West Oncology Group (SWOG) phase III trial compared bolus versus infusional administration of doxorubicin plus DTIC and reported no differences in overall response (17% in both arms) or complete responses (5% in both arms). Additionally, there was no difference in the median survival, 10.5 months in both groups (Zalupski et al., 1991). Adding cyclophosphamide to the basic duo was reported to raise the response rate to 56% (Blum et al., 1980) and this was confirmed by a randomized trial (Baker, 1987). Comparisons have shown that the addition of less active drugs necessitate lower doses of doxorubicin and , accordingly, reduces overall effectiveness (Cruz et al., 1979; Schoenfeld et al., 1982). Adding ifosfamide seems to be clearly beneficial as report by Blum et al, (1993) and Schutte et al, (1993).

The ECOG conducted a three-arm trial comparing doxorubicin alone, doxorubicin plus ifosfamide, and mitomycin plus doxorubicin plus cisplatin. Objective tumour

regression occurred more frequently in the combination arms than in the single-agent arm (20% with doxorubicin alone, 34% in doxorubicin plus ifosfamide, and 32% in the mitomycin plus doxorubicin plus cisplatin arm); However, the combination regimens resulted insignificantly greater myelosuppression. e.g, 80% of the doxorubicin/ifosfamide group had grade 3 or greater myelosuppression. Most notably, no significant survival differences were observed between the three treatment regimens (Edmonson et al., 1993).

The extensively utilized CYVADIC regimen have evolved from the sequential trials sponsored by SWOG (Gottlieh et al., 1975). Greenall et al., (1986) reviewed studies reporting response rates of 15-60%, the average being 41%. Noteworthy is that up to 15% of the responses have been scored as complete. The median response was longer (13 months) and 21% of complete responses described by Yap et al,(1986) have also reviewed alternative regimens. A popular regimen adds ifosfamide to Adriamycin and DTIC (Elias and Antman, 1986). A combination of ifosfamide with mesna, doxorubicin and DTIC has resulted in response rates in measurable metastatic sarcomas as high as 47% with complete response rates as high as 10% (Chang et al., 1989; Elias et al., 1989). Another protocol described activity with DTIC and cisplatin in pretreated patients (Piver et al., 1986).

Dose intensity may be extremely important as described by Zanineli et al., (1993). Very high-dose ifosfamide had been used by Rosen et al, (1994) with high response rates despite some toxicity. Higher-dose therapy with standard agents (Bodey, 1981) may require special supportive care such as bone marrow transplantation (Kesinger et

al., 1994), but may offer a chance for higher complete response rates and longer response duration. There has been a considerable interest focused on maintaining dose intensity of chemotherapy using colony stimulating factors to alleviate myelosuppression. Granulocyte/macrophage colony stimulating factor (GM-CSF) has been used with a variety of regimens to help maintain dose intensification. In a few studies this has resulted in improved response rates (Mertens and Bramwell, 1993). Even higher dose chemotherapy, as used at M.D. Anderson, seems to result in higher response rates (59-69%) (Pisters et al., 1997; Patel et al., 1998).

PULMONARY SURGERY FOR METASTATIC DISEASE

Distant metastases usually present 2 years after the initial diagnosis. Patients with high-grade sarcomas have a higher risk of developing distant disease compared with low-grade extremity sarcomas (Donohue et al., 1988). For patients with extremity tumours, metastatic disease most frequently appears in the lungs (Gadd et al., 1993). Evidence is now available that resection of pulmonary metastases may be worthwhile in selected patients. Factors such as disease-free interval (time elapsing between resection of the primary tumour and appearance of pulmonary metastases) and tumour doubling time (calculated from measurements of the diameter of pulmonary nodules seen on serial chest X-rays) influence prognosis. The following criteria for selection of patients for surgery have been suggested (Joseph, 1974):

- Control of the primary tumour has been achieved;
- There is no evidence of extrapulmonary metastases;

- Pulmonary lesions should be resectable, multiple lesions are not a contraindication but if extensive multiple resections are necessary the failure rate rises;
- The patient should be fit for surgery as regards general condition and respiratory status;
- Tumour doubling time should be atleast 40 days,

Complete resection of pulmonary lesions is possible in some patients who have limited disease and adequate pulmonary function.

MATERIALS AND METHODS

MATERIALS AND METHODS

Though the soft tissue sarcomas rare tumours, In our institution,we are treating a significant number of soft tissue sarcomas, Since our hospital has well established oncology department. The study design is a retrospective analysis about "**Limb salvage in extremity soft tissue sarcomas**". This Study was conducted in Government Royapettah Hospital . Study period was 5 years duration between (Jan' 2001 – Dec' 2005). All the extremity soft tissue sarcomas patients who were attending our outpatient department and underwent treatment included in this study. All the age group of patients were taken for this study. Sex prevalence has been calculated.

Any soft tissue swelling which were > 5cms, or any suspicion of malignant features like sudden increase in size , adherent with a underlying fascia, recurrence after excision and any evidence of metastasis, these patients were subjected to Trucut Biopsy after thorough clinical examination.

For superficial swelling, that are smaller than 5cm, particularly those were suspected for malignancy, Excisional biopsy was done. For more than 5cm sized and deep seated either Trucut biopsy or Incisional biopsy were done . Trucut biopsy was done in such way that the needle punctured site and track of the needle pathway were included in definitive surgery, If it could be proved to be as soft tissue sarcoma.

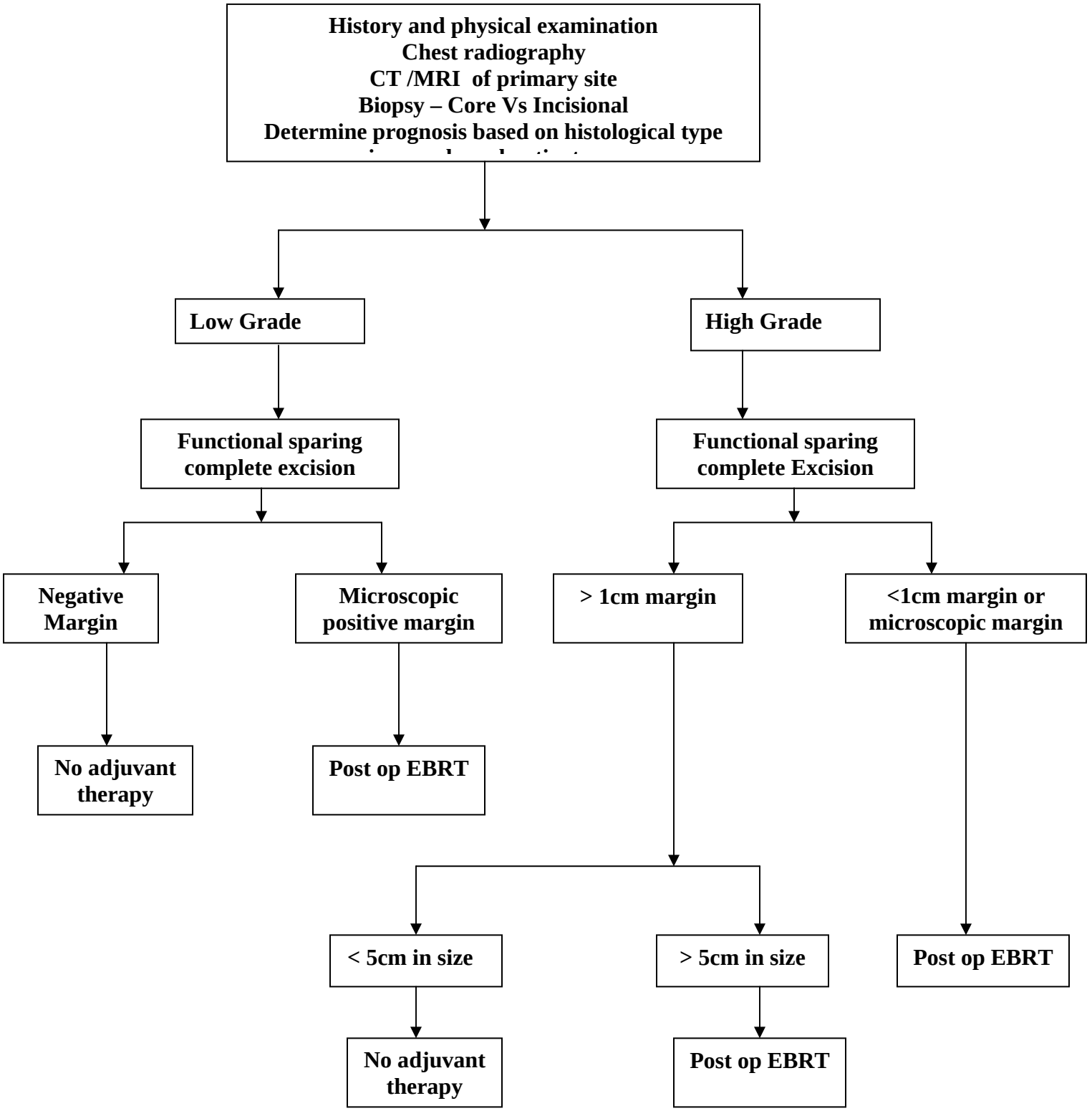
Incisional biopsy - the incision was made along the long axis of the affected limb, within the tumour limit and was planned to include the biopsy scar in following

definitive surgical procedure. After taking adequate tissue for HPE, perfect hemostasis was achieved and no drain was kept.

Role of FNAC in diagnosing sarcomas are limited. FNAC will be done for suspected recurrence after surgery and to diagnose sarcomas which are situated in deeper plane in trunk and abdomen. This is because the tissue material that we are getting through FNAC is inadequate to grade and subtype the tumour, which is essential in deciding its prognosis and choosing adjuvant therapy. After histological confirmation , staging work up will be done by using conventional radiographs, CT and MRI for local disease , And radiographs and CT for distant metastasis.

After complete evaluation, the extremities soft tissue sarcomas with distal metastasis, inoperable lesions, medically unfit or not willing for surgery were referred to palliative treatment. Other patients were taken for surgery after proper counselling.

Department protocol for management of extremity soft tissue sarcomas:



EBRT – External Beam Radiotherapy

Guidelines for Limb sparing resection:

1. No major neurovascular tumour involvement.
2. Wide resection of affected part with a normal cuff tissues in all direction.
3. En bloc removal of all previous biopsy sites and potentially contaminated tissue
4. Adequate motor reconstruction.

Contraindications for limp sparing surgery

1. major neurovascular involvement
2. pathologic fracture
3. inappropriate biopsy site
4. Gross infection
5. skeletal immaturity
6. extensive muscle involvement

Limb sparing surgery were done to two group of patients. The first group were those who underwent primary surgery in our institution. The second group of patients to whom the primary surgery was done elsewhere and referred with local recurrence. They were restaged and those with distant metastasis were referred for palliative treatment. Other were underwent surgical treatment either limb sparing procedure or amputation which were decided by above mentioned factors . Those patients who were not fulfilling the above guidelines , underwent amputation. All the groups were studied and registered individually

Follow up:

First year	every month
Second year	every second month
Third year	every third month
4&5 th years	every six month
6 th year onward	annually

On every follow up:

1. Patient had full clinical examination

(On basis of suspicion of recurrence or metastasis or otherwise annually)

2. X-ray chest PA view
3. Radiograph of part concerned if necessary
4. For suspected lesion in chest, CT chest was suggested
5. For suspected local recurrence Trucut Biopsy / FNAC ± MRI was done

Death rate : Documented death rate has been taken for this study. But many patients lost their follow up. Here, the projected death may not reflect the actual death rate.

RESULTS& OBSERVATION

RESULTS AND OBSERVATION

Between Jan' 2001 – Dec' 2005, about 107 patients of extremity soft tissue sarcomas were treated in our institution. After metastatic work up, about 41 patients were found to be with distal metastasis and two patients were at inoperable stage due to extensive soft tissue involvement at critical anatomical site. These 43 patients were referred for palliative chemoradiotherapy. Others underwent multimodality treatment.

OBSERVATION

I. SEX DISTRIBUTION

Sixty cases were male and forty seven cases were female.

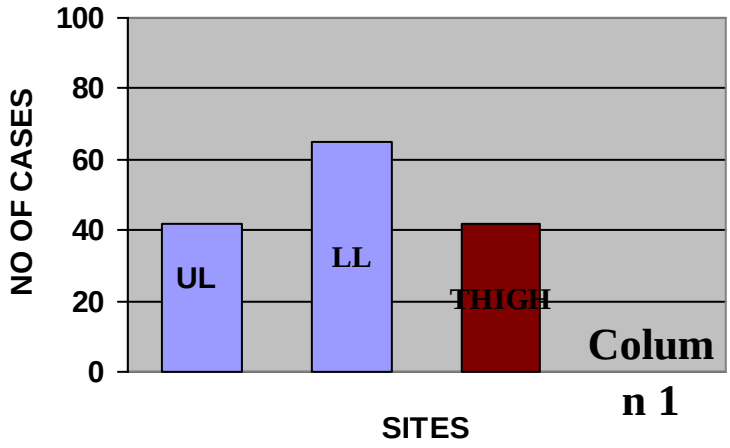
Male : Female ratio is 1.3 : 1.

II. ANATOMICAL DISTRIBUTION:

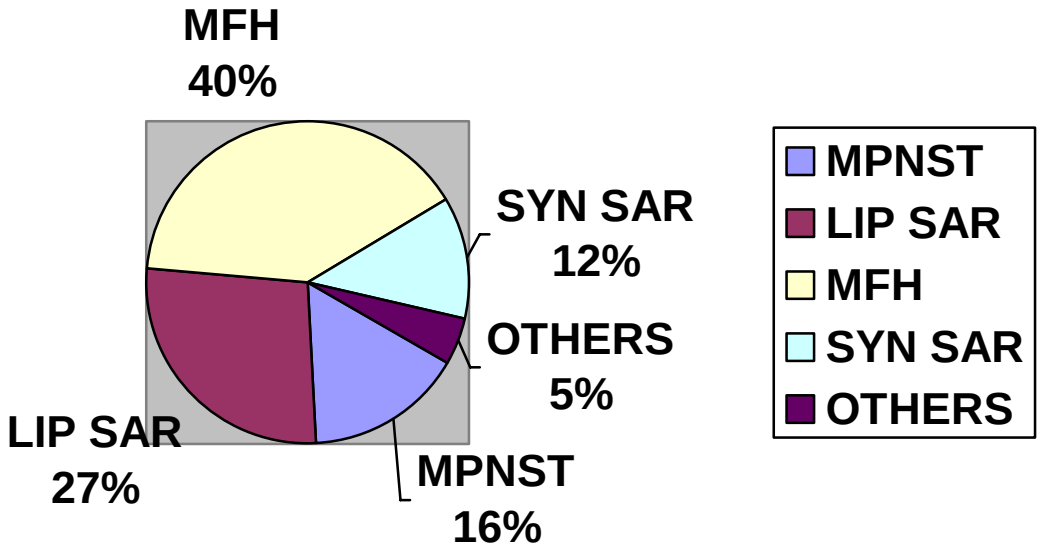
Out of 107 cases, Upper limb – 42 cases (39.3%)

Lower limb – 65 cases (60.7%)

Thigh alone – 42 cases (39.3%)



III HISTOLOGICAL TYPES OF SOFT TISSUE SARCOMAS



MPNST-Malignant peripheral nerve sheath tumour,LIP SAR-Liposarcoma

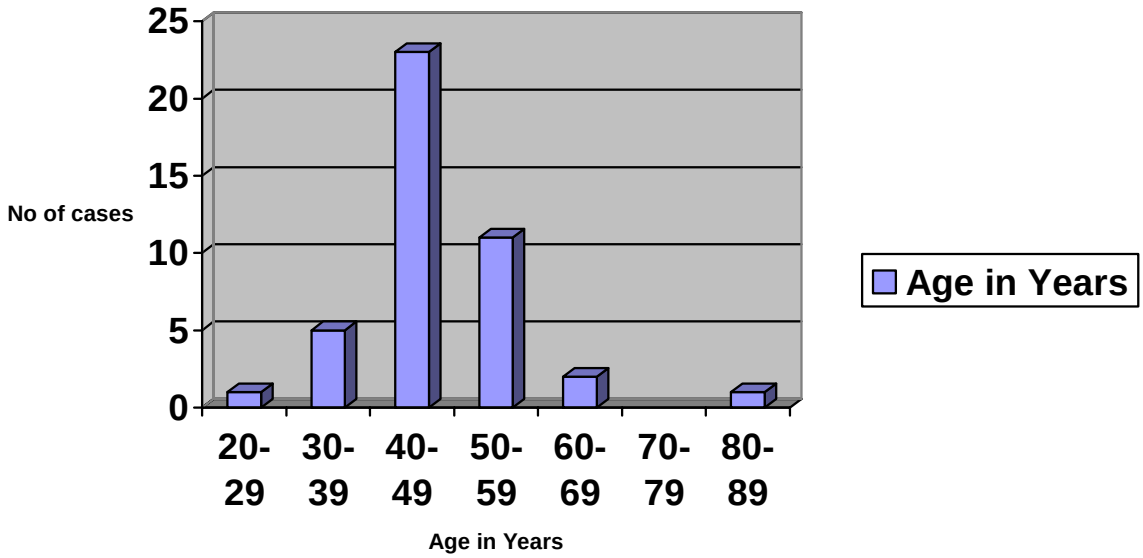
MFH-Malignant fibrous histiocytoma,SYN SAR-Synovial sarcoma.

Malignant fibrous histiocytoma	n - 43 (40.2%)
Liposarcomas	n - 29 (27.1%)
MPNST	n - 17 (15.9%)
Synovial sarcoma	n - 13 (12.1%)
Others	n - 5 (4.7%)

IV AGE DISTRIBUTION

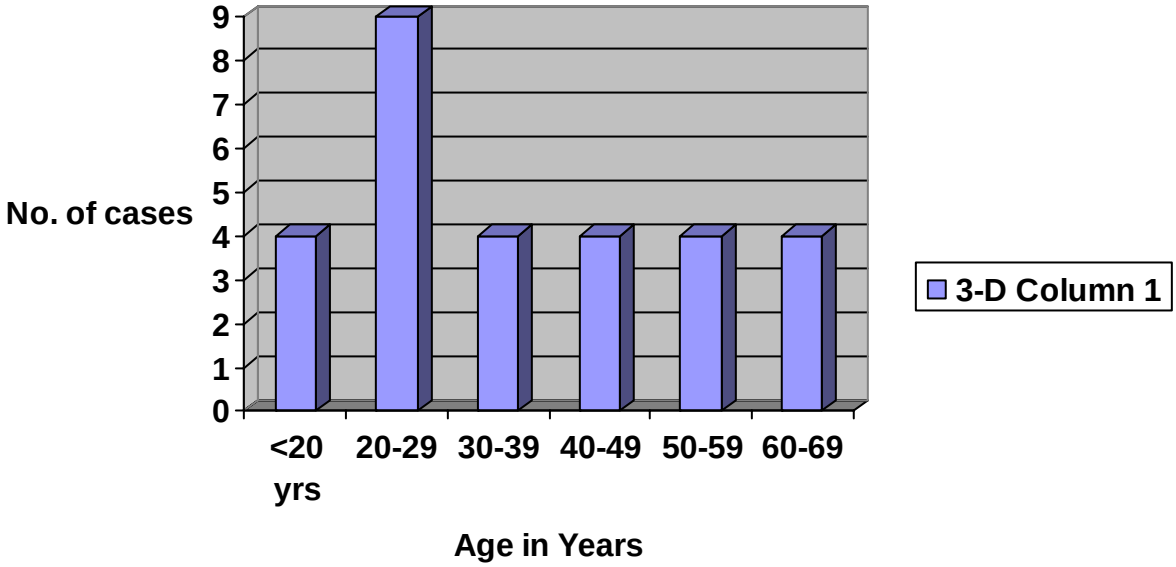
1. Malignant fibrous histiocytoma n - 43

Age in years	Cases
20-29	**1**
30-39	**5**
40-49	**2**
50-59	**11**
60-69	**2**
70-79	**Nil**
80-89	**1**



2. Liposarcoma n – 29 cases

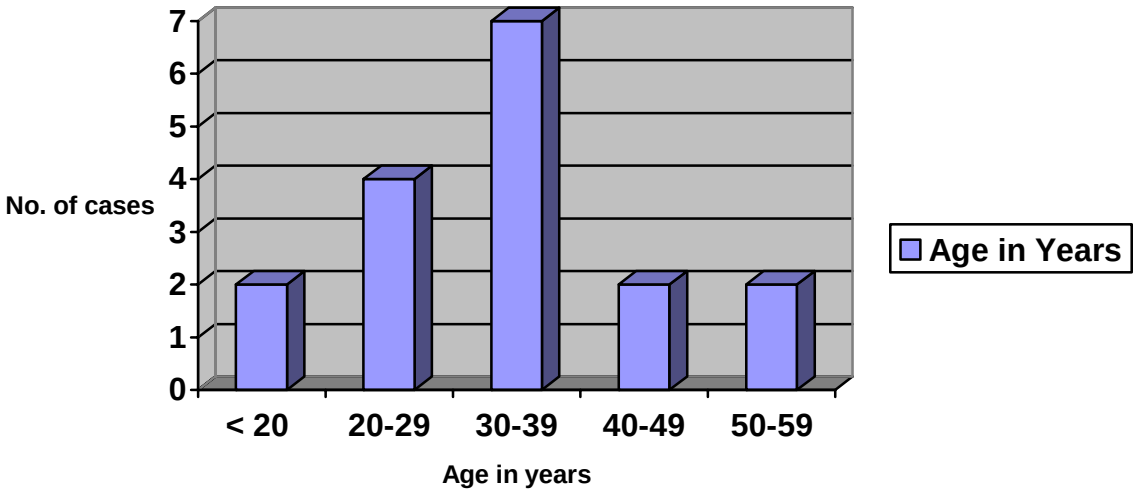
< 20 yrs	**4**
20-29	**9**
30-39	**4**
40-49	**4**
50-59	**4**
60-69	**4**



3. Malignant Peripheral Nerve Sheath Tumours

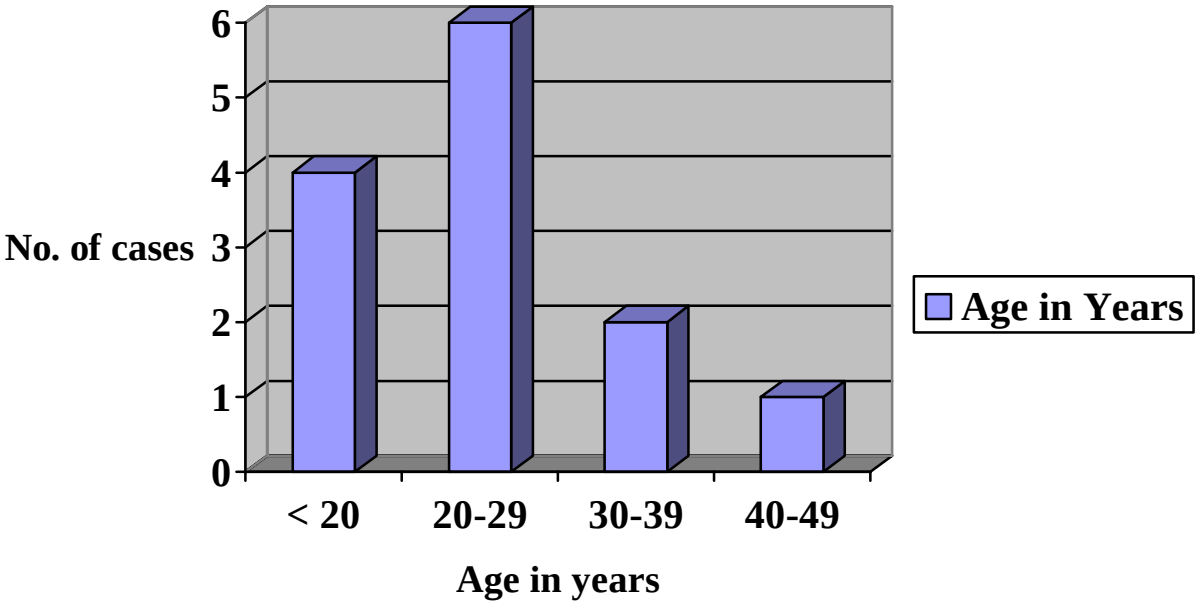
n – 17 cases

< 20 yrs	**2**
20-29	**4**
30-39	**7**
40-49	**2**
50-59	**2**



4.Synovial Sarcoma n -13 cases

< 20 yrs	**4**
20-29	**9**
30-39	**4**
40-49	**4**



6.Others n – 5 cases.

IV TREATMEMT STATISTICS

Total number of patients underwent surgery	**Limb sparing surgery**		Amputation
	Primary surgery	**Reresection For local recurrence**	
64	**25**	**31**	8

a. Twenty eight cases were underwent primary surgical treatment in **our institution .**

Wide local excision	-	20 cases (71.4%)
Compartmental resection	-	4 cases
Rotation plasty	-	1 case
Total limb sparing surgery	-	25 cases (89.2%)
Amputation	-	3 cases (10.8%)

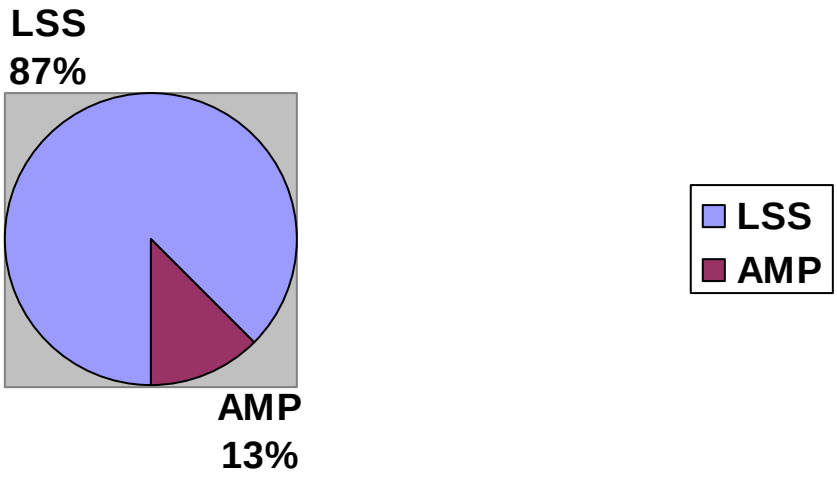
b. Second surgery for **recurrence Soft tissue sarcoma** primarily operated **elsewhere**

Total no. of cases	-	36
Wide local excision	-	25(69.4%)
Compartmental resection	-	6
Total (limb sparing surgery)	-	31 (86.1%)

Amputation - 5 (13.9%)

Over all Limb sparing surgery - 56 (87.5%)

Amputation - 8 (12.5%)



LSS- Limb sparing surgery ***AMP- Amputation***

V RECURRENCE RATE

After surgical treatment six patients developed local recurrence of which two(7.1%) from the group who underwent primary resection, four(11.1%) from the group who underwent re-resection for local recurrence.

All the local recurrence were in patient who underwent limb sparing surgery. No amputee developed local recurrence. Local recurrence rate is 9.4%.

Local recurrence	After primary surgery in	After re-resection

	our institute	(first surgery elsewhere)
6(9.4%)	*2(7.1%)*	*4(11.1%)*

METASTASIS

Ten patients developed distant metastasis. All were seen in lung (100%). Seven patients who underwent limb sparing surgery and three patient who underwent amputation developed pulmonary metastasis. Two patients developed both local recurrence and pulmonary metastasis simultaneously. All metastasis and local recurrence developed within 2 years of initial diagnosis. All were the high grade tumour.

DEATH

Eleven patients has been document as died in hospital due to soft tissue sarcoma in 5 years follow up with mean follow up of 29 months of which 9 patients were with distal metastasis (81.8%). Many patients lost for follow up.

DISCUSSION

DISCUSSION

Soft tissue sarcomas are rare tumour. Since our institution is a tertiary centre and has well established oncology department, we are treating a significant number of soft tissue sarcomas. An analysis of population based data from Connectiout suggest increased incidence in both men and women, with a greater increase in women. This study shows increase number of soft tissue sarcomas in both male and female with minimal male preponderance. (M:F ration 1.3:1).

In MSKCC series shows mean age of soft tissue sarcoma occurrence as 51 years with range of 16-93 years and most of them are seen in sixth decade. In this review shows that age distribution range of 13-80 years with mean age of 39.1 years. Majority of cases were seen in fifth decade (40-49) years, especially malignant fibrous histiocytoma, the most common soft tissue sarcoma which is reported as more common in seventh decade. But in this review, MFH is common in fifth decade.

Lawrence et al, in his series found as soft tissue sarcomas in upper extremity as 30%, and lower extremity as 70%. In this study, we found upper extremity soft tissue

sarcoma were about 39.3% and lower extremity as 60.7% Thigh is most common site for soft tissue sarcoma. This has been supported by this study which found about 39.3% of extremity soft tissue sarcoma seen in thigh region.

In a study conducted in MCH U.S(1994) showed malignant fibrous histiocytoma as most common soft tissue sarcoma (21.9%) followed by liposarcoma (16.0%) and MPNST(9.6%). In this review shows most common extremity soft tissue sarcoma is MFH (40.2%) followed by LPS (27.1%). Surprisingly MPNST accounted as (15.9%). By comparing both studies there is increased incidence of MFH and MPNST in our region. Synovial sarcoma is common among young adult. In this study shows most of the synovial sarcomas were seen in second and third decade (76.9%).

Study series	Total number of patients operated	Limbsparing surgery	Amputations
Maghert et al	413	388 (94%)	25 (6%)
Sluga et al	130	116(89.2%)	14(10.8%)
Our study results	**64**	**56 (87.5%)**	**8 (12.5%)**

Before 1991, 90% of extremity soft tissue sarcoma were treated by amputation. After 1990, with better knowledge about biology of soft tissue sarcoma, availability of

imaging modality, multimodality of treatment, improvement limb reconstruction technique and improved rehabilitation facilities, now about 90% of extremity soft tissue sarcoma are treated by limb salvage procedure. In this review study., out of 28 newly diagnosed soft tissue sarcoma, limb sparing surgery was done in 25 cases (89.2%). Where as amputation was done in 3 cases (10.7%). In recurrence soft tissue sarcoma (36 cases) limb sparing surgery was achieved in 31 cases (86.1%). Amputation was done in 5 cases (13.8%). The minimal increase in amputation in the group with Re-resection were due to unplanned previous surgery like unknowingly tumour was removed as benign or improper biopsy procedure or intralesional / intracapuslar excision.

This led to difficulty in planning for second limb sparing procedure. In spite of this overall limb salvage rate of 87.5% was achieved. This study confirms the influence of surgical margin achieved in development of local recurrence and stress the importance of proper planning of primary surgery to minimize local recurrence .

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STUDY SERIES	Wirbel et al	Enneking et al	Sluga et al	**Our study results**
Local recurrence after limb sparing surgery.	9.3 %	5%	2.3%	**9.4%**

Wirbal et al, his series showed local recurrence rate of 9.3% after wide local excision with margin negative specimen and 28.5% after marginal margin resection. Simon and Enneking et al, in their series, found 5% local recurrence after negative margin and 89% of local recurrence after positive margin. In this study, After surgical treatment, six patients (9.4%) developed local recurrence of which two (9.1%) patients after primary resection. Four(11%) patients after re-resection developed local recurrence. This definitely shows the increase in local recurrence in re-resection group was due to unplanned first surgery which made difficulty in second limb sparing surgery. Moreover 5 out of 6 local recurrence were margin positive cases (83.3%).

Lung is the commonest site for distal metastasis extremity sarcomas. In this study ten patients developed distal metastasis All of them were pulmonary metastasis (100%). A study about extremity soft tissue sarcoma showed isolated lung metastasis as 70%, multiple metastasis as 5%. More than 60% of metastasis are found within 2 years. In this study, All lung metastasis were found with mean follow up of 29 months. Three metastasis were seen after amputation., which shows local control of soft tissue sarcoma does not alter the metastatic rate. By this, it is evident that local control of disease and metastasis are two separate entity which are individually determined by stage of the tumours. Achieving complete local control of disease will not alter the chance of distant metastasis.

Documented death of 11 cases were reported of which nine were with metastatic disease. It is evident as metastasis shorten the overall survival rate. Since many patients lost their follow up , this death rate is not reflecting the real picture of overall survival in soft tissue sarcomas of extremity after multimodality treatment.

CONCLUSION

CONCLUSION

This retrospective review on "Limb salvage in extremity soft tissue sacomas" for five years period shows:

- Occurrence of limb soft tissue sarcomas has shifted one decade Iin our region earlier than literature evidence. More number of soft tissue sarcoma seen in younger age with minimal male preponderance
- Histologically, malignant fibrous histiocytoma is the most common extremity soft tissue sarcoma, followed by liposarcoma and occurrence of MPNST seems to be increased in our region
- Anatomically lower limb is common site, especially thigh is the most common site for limb sarcomas
- Due to better knowledge about soft tissue sarcoma, availability of imaging modality and multimodality treatment and improvement in limb

reconstruction technique, Limb salvage procedure is achievable in about 90% of extremity soft tissue sarcomas.

- Local recurrence after limb sparing surgery is only 9.4% and shows success of limb sparing technique.
- Local recurrence are higher in re-resection group due to unplanned previous surgery which make the second surgery difficult to achieve limb salvage.
- First surgery is the best surgery as far as limb salvage and local control of disease concerned.
- Ideally biopsy should be done by the same surgeon who is going to do definitive surgical procedure
- Distant metastatic potential is not influenced by successful local control
- Local recurrence and distant metastasis are determined by tumour size, grade and plane of swelling as individual factor
- .Lung is the most commonest site of distal metastasis for extremities soft tissue sarcoma.

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ANNEXURE

PROFORMA

a. Particulars of patient

i. Name

ii. Age

iii. Sex

iv. Occupation

v. Religion

vi. Residence

vii. In patient No:

viii. Date of Admission

ix. Date of surgery

x. Date of discharge

b. Complaints and Duration

i. Swelling

ii. Ulceration

iii. Pain

iv. Numbness

v. Paresthesia

vi. Fever

vii. Fainting

viii. Appetite

ix. Loss of weight

x. Alteration in function of limb

xi. Cough

xii. Haemoptysis

xiii. Bone pain

xiv. Jaundice

xv. Paralysis

xvi. Similar swelling

xvii. Conclusion

xviii. Fracture

III Past history

1. Tuberculosis
2. Diabetes mellitus
3. Hypertension
4. Jaundice
5. Burns
6. Radiation
7. Trauma
8. Previous surgery
9. any other malignancy

IV Personal History

1. Dietary Habits
2. Smoking
3. Alcoholism

V Occupational History

Employment in dye industry

VI Family History

Similar illness in any other members of I relatives

VII Physical Examination

1. **General Examination**
    a. Nutritional status
    b. Anaemia
    c. Jaundice
    d. Pedal edema
    e. Dyspnoea
    f. Clubbing
    g. Generalized / significant lymph adenopathy
2. **Local examination**
    a. Location of tumour in relation to known anatomical land marks
    b. Characteristics of tumour
    c. Involvement of adjacent structure
    d. Confinement to anatomical compartment
    e. Distal neurovascular deficit
    f. Near by joint movement
3. **Nodal status**
    a. Not involved
    b. Involved –Ipsilateral / Bilateral, features of lymph nodes
4. **Distant metastasis**
5. **Clinical diagnosis (includes clinical staging)**

VIII Histological features : type / subtype grade

IX Investigations

X Treatment planned

XI Outcome

XII Remarks

XIII Follow up

MASTER CHART

NEW CASES – UPPER LIMB SOFT TISSUE SARCOMAS

S.No	NAME	AGE/SEX	I.P No	ANATMOMICAL SITE	HISTOLOGICAL TYPE/ GRADE	SURGERY	REMARKS
1	RAJESHKUMAR	28/M	808018	RT. SCAPULAR REGION	LIPOSARCOMA, LG	WLE	-
2	ELANGO	45/M	801841	LT SHOULDER AREA	MFH, HG	WLE	-

3	ARIVALAGAN	50/M	816443	RT FOREARM	LIPOSARCOMA LG	WLE	-
4	SHUBA	27/F	813081	LT ELBOW	SYNOVIAL SARCOMA	AE AMPUTATION	-
5	MURUGAN	41/M	813776	RT FOREARM	MFH, HG	AE AMPUTATION	M/D
6	MUSTAFA	80/M	783116	RT ELBOW REGION	MFH	WLE	-
7	GNANAROSE	27/F	726478	RT FOREARM	DFSP, LG	WLE	R
8	MANOHARAN	42/M	728127	RT ARM	MFH, HG	WLE	-
9	JAYALAKSHMI	40/F	710487	LT ARM	MFH, HG	WLE	-
10	PALANI	34/M	719263	RT ARM	MPNST	WLE WITH ROTATION PLASTY	-
11	KUMAR	47/F	719667	LT. FOREARM	SYNOVIAL SARCOMA	WLE	-
12	RADHIKA	21/F	716465	RT. FOREARM	MPNST	WLE	-

M-Metastasis, R-Recurrence, D-Died.

NEW CASES LOWER LIMB

S.No	NAME	AGE/SEX	I.P No	ANATMOMICAL SITE	HISTOLOGICAL TYPE/ GRADE	SURGERY	REMARKS
1	SAMPATH	30/M	778021	RT. SILIAC REGION	MPNST, HG	RT. INTERNAL HEMIPELVECTOMY	-
2	MURALI	15/M	774057	LT THIGH	LIPOSARCOMA, HG	WLE	M
3	SELVAKUMAR	15/M	775264	LT THIGH	SYNOVIAL LG	WLE	-

4	CHITTU	40/F	789391	RT THIGH	MFH, HG	WLE	-
5	ROYAMMAL	55/F	787956	LT THIGH	MFH, HG	WLE	-
6	PADMAVATHY	40/F	794205	LT THIGH	MPNST, HG	WLE	-
7	MANI	56/M	797454	LT THIGH	MFH, HG	HAMSTRING COMPARTMENTAL RESECTION	-
8	SRINIVASAN	36/M	786791	LT THIGH	MPNST, HG	HAMSTRING C.R	M/D
9	MUNIAMMAL	39/M	726610	LT LEG	SYNOVIAL SARCOMA	LT AK AMPUTATION	-
10	MARIAPPAN	21/M	742111	RT THIGH	LIPOSARCOMA, LG	WLE	-
11	SHAKTHI	36/M	752105	LT THIGH	LIPOSARCOMA, LG	WLE	-
12	RUKMANI	56/F	756341	LT THIGH	MFH, HG	WLE	R
13	MUTHULAKSHMI	55/F	753165	LT THIGH	MFH, HG	WLE	-
14	POONGODI	45/F	759635	RT THIGH	MFH, HG	WLE	-
15	GANDHIMATHI	36/F	766849	LT LEG	SYNOVIALMFH, HG SARCOMA, HG	WLE	-
16	RAJAN	62/M	776526	RT THIGH	LIPOSARCOMA, HG	ADDUCTOR C.R	-

M-Metastasis, R-Recurrence, D-Died.

RECURRENCE SOFT TISSUE SARCOMAS – UPPER LIMB

S.No	NAME	AGE/SEX	I.P No	ANATMOMICAL SITE	HISTOLOGICAL TYPE/ GRADE	SURGERY	REMARKS
1	SHANTHI	40/F	821745	RT ARM	MFH, HG	RT SHOULDER DISSARTICULATION	M/D
2	DHANALAKSHMI	45/F	821898	LT FOREARM	MFH, HG	AE AMPUTATION	-
3	PURUSHOTHAMAN	27/M	824549	LT ELBOW AREA	MPNST, HG	WLE	-

4	DHANALKSHMI	31/F	826617	LT ARM	MPNST, HG	WLE	-
5	BOOPATHY	34/F	89220	RT FOREARM	MPNST, HG	WLE	M/D
6	STELLAMANY	30/F	779394	RT ARM	LIPOSARCOMA, LG	WLE	-
7	MUNIAMAL	55/F	784074	RT ELBOW REGION	MFH, HG	WLE	-
8	GANGAN	53/M	782128	LT ARM	LIPOSARCOMA, LG	WLE	-
9	JAYALAKSHMI	45/M	772979	RT FORE ARM	MFH, HG	WLE	-
10	RAMANI	17/F	781031	RT ARM	SYNOVIAL SARCOMA	WLE	-
11	DEVI	40/F	723141	RT ARM	LIPOSARCOMA, HG	WLE	-
12	MURUGESAN	45/M	720385	LT ELBOW AREA	MPNST, HG	WLE	-
13	BALARAMAN	50/M	763953	LT SCAPULAR REGION	FIBROSARCOMA, HG	LT SCAPULECTOMY	-
14	VIJAYAKUMAR	23/M	785040	RT FORE ARM	LIPOSARCOMA, LG	WLE	-

M-Metastasis, R-Recurrence, D-Died.

LOWER LIMB

S.No	NAME	AGE/SEX	I.P No	ANATMOMICAL SITE	HISTOLOGICAL TYPE/ GRADE	SURGERY	REMARKS
1	SAMRUTHI	30/F	714355	SACROGUTEAL REGION	LIPOSARCOMA, LG	WLE	-
2	GOVINDAMMAL	51/F	818752	LT THIGH	MFH, HG	WLE	-
3	ANJALAGAN	47/F	821643	RT THIGH	MFH, HG	ADD, C.R	R
4	SAMPATH	34/M	822587	LT THIGH	MFH, HG	LT H.P DISARTICULATION	-
5	KANJANA	19/F	827598	LT THIGH	ANGIOSARCOMA	WLE VASCULAR RECONSTRUCTION	-

6	LALITHA	19/F	827598	LT THIGH	LIPOSARCOMA, HG	ADDN C.R	-
7	RAMAPRIYA	28/F	814389	RT POPLITHEAL FOSSA	LIPOSARCOMA, HG	WLE	M/D
8	ANITHA	21/F	815745	LT THIGH	SYNOVIAL SARCOMA, HG	WLE WITH INGUINAL BLOCK DISSECTION	-
9	RANJITH	15/M	765849	RT GLUTEAL AREA	LIPOSARCOMA, HG	WLE	D
10	VADIVEL	18/F	762537	LT THIGH	MPNST, HG	WLE	-
11	PATCHIYAPPAN	29/M	772344	RT LEG	MPNST, HG	WLE	M/D
12	MURALI	16/M	724057	RT THIGH	LIPOSARCOMA. LG	HAMS. C.R	-
13	KAMALA	55/F	762102	RT THIGH	MFH, HG	WLE	-
14	KALIMUTHU	45/F	754196	RT LEG	MFH, HG	RT AE AMPUTATION	-
15	SUKUMAR	25/M	762994	LT LEG	SYNOVIAL SARCOMA, HG	WLE	-
16	BALAJI	18/M	760443	LT FOOT	SYNOVIAL SARCOMA, HG	SYMES AMPUTATION	-
17	SATHESH	22/M	760669	LT POPLITIEAL FOSCA	MFH, HG	WLE	M/D
18	BHARATHI	25/F	737551	LT LEG	MPNST, HG	WLE	-
19	VIJAYALAKSHMI	32/F	737742	LT LEG	MPNST, HG	POSTERIOR C.R	R/M/D
20	DEVI	40/F	723141	RT THIGH	MFH, HG	ANTERIOR C.R	-
21	NAGARAJ	35/M	720654	RT LEG	SYNOVIAL SARCOMA, HG	WLE	-
22	MURUGESAN	45/M	720585	LT THIGH	MFH, HG	C.R	-

LG- low grade, HG- high grade, WLE- wide local excision, CR- compartmental resection. M-Metastasis, R-Recurrence, D-Died.

SOFT TISUE SARCOMAS WITH DISTANT METASTASIS

S.No	NAME	AGE/SEX	I.P No	ANATMOMICAL SITE	HISTOLOGICAL TYPE/ GRADE	METASTATIC SITE
1	PRAKASH	37/M	701928	LT THIGH	LIPOSARCOMA, LG	LUNG
2	SASIKALA	59/F	703552	LT GROIN	MPNST, HG	LUNG
3	RUKMANI	48/F	704604	RT THIGH	MFH, HG	LUNG
4	RADHIKA	40/M	706465	RT THIGH	MFH, HG	LUNG
5	PURUSOTHAMAN	56/F	708948	LT ARM	LIPOSARCOMA, HG	LUNG

6	BAKYALAKSHMI	55/F	708862	LT ARM	MFH, HG	LUNG
7	BADHAR	25/F	708860	RT ARM	MPNDT	LUNG
8	MANAVALAN	56/M	712609	RT THIGH	MFH, HG	LUNG/SKULL BONE
9	PLANIESHWAR	60/F	712884	RT THIGH	LIPOSARCOMA, HG	LUNG
10	SHAHANA	28/F	714624	RT GLUTEAL REGION	LIPOSARCOMA, HG	LUNG
11	IYYANAR	62/M	724530	RT GLUTEAL REGION	MFH, HG	LUNG
12	MANIKANDAN	43/M	724896	RT THIGH	MFH, HG	LUNG
13	MANAVALAN	38/M	726849	RT THIGH	MFH, HG	LUNG
14	VALARMATHY	32/F	728721	RT FOREARM	LIPOSARCOMA, HG	LUNG
15	SIVAKUMAR	40/M	791346	RT THIGH	MFH, HG	LUNG
16	JAYALAKSHMI	52/F	72032	LT FOREARM	MFH, HG	LUNG
17	ROSSIAMMAL	60/F	751278	LT THIGH	LIPOSARCOMA, HG	LUNG
18	PARAMESWARI	43/F	751014	LT LEG	LIPOSARCOMA, HG	LUNG
19	MURUGAN	48/F	752285	RT SHOULDER AREA	MFH, HG	LUNG
20	SUKUMAR	47/M	752994	RT GLUTEAL REGION	LIPOSARCOMA, HG	LUNG/LIVER
21	SENTHILKUMAR	34/M	754219	RT LEG	MFH, HG	LUNG
22	KAVIKARASI	55/F	756952	LT FOREARM	ANGIOSARCOMA, HG	LUNG & LIVER
23	RAJAMANICKAM	65/M	761391	RT THIGH	MGH, HG	LUNG
24	THUKKARAM	58/M	762915	LT GROIN	LIPOSARCOMA, HG	INOPERABLE
25	NALLATHAMBI	42/M	760349	RT ARM	LIPOSARCOMA, HG	LUNG
26	ANNAMMAL	45/F	763596	RT THIGH	LIPOSARCOMA, HG	LUNG
27	VADIVEL	45/M	762537	LT ELBOW AREA	MFH, HG	LUNG
28	LATHA	43/F	759213	LT THIGH	MFH, HG	LUNG
29	PATCHIYAPPAN	38/M	772344	LT LEG	MFH, HG	LUNG
30	KARTHIK	24/M	784401	RT ELBOW	SYNOVIAL SARCOMA, HG	LUNG
31	DIVYA	29/FF	786434	RT LEG	LIPOSARCOMA, HG	LUNG
32	PRAKASH	39/M	783094	LT FOREARM	MFH, HG	LUNG
33	SHINEE	29/F	791070	LT THIGH	LIPOSARCOMA, HG	LUNG/SPINE
34	LINGAM	60/M	798932	RT ARM	MFH, HG	LUNG
35	BALAMURUGAN	48/M	796903	RT THIGH	MFH, HG	LUNG

36	**SHANKAR**	**39/M**	**795041**	**LT ELBOW**	**SYNOVIAL SARCOMA, HG**	**LUNG**
37	**MOHAN**	**52/M**	**800492**	**LT THIGH**	**MPNST, HG**	**INOPERABLE**
38	**RAMESH**	**43/M**	**802859**	**RT LEG**	**MFH, HG**	**LUNG**
39	**VIJAY**	**18/M**	**805070**	**RT THIGH**	**SYNOVIAL SARCOMA, HG**	**LUNG**
40	**BALARAMAN**	**39/M**	**813953**	**LT THIGH**	**MFH, HG**	**LUNG**
41	**ALAGANBAN**	**48/M**	**817548**	**LT ARM**	**MFH, HG**	**LUNG**
42	**KANNAN**	**21/M**	**822003**	**LT LEG**	**SYNOVIAL SARCOMA**	**LUNG**
43	**SATHIYA**	**27/F**	**823605**	**RT SHOULDER REGION**	**MPNST, HG**	**LUNG**

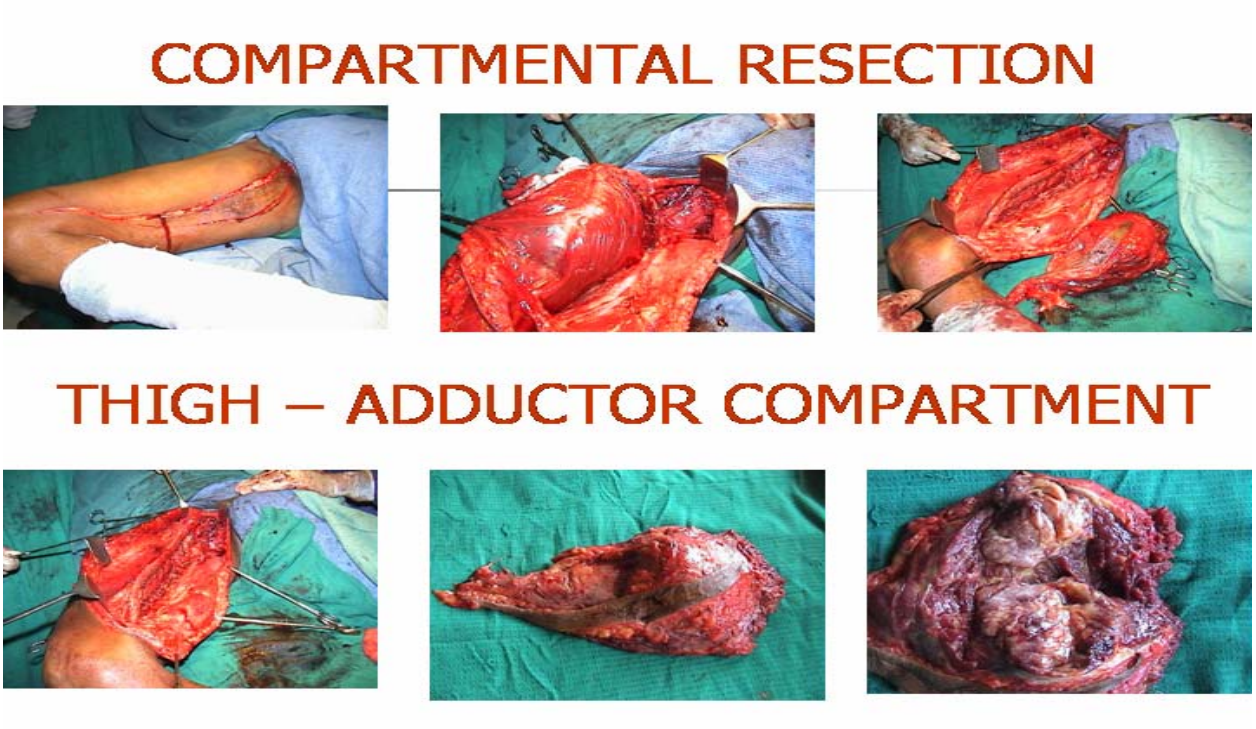


Figure 3 illustrate , A 58 yrs old man , a case of malignt fibrous histiocytoma underwent adductor compartmental resection.

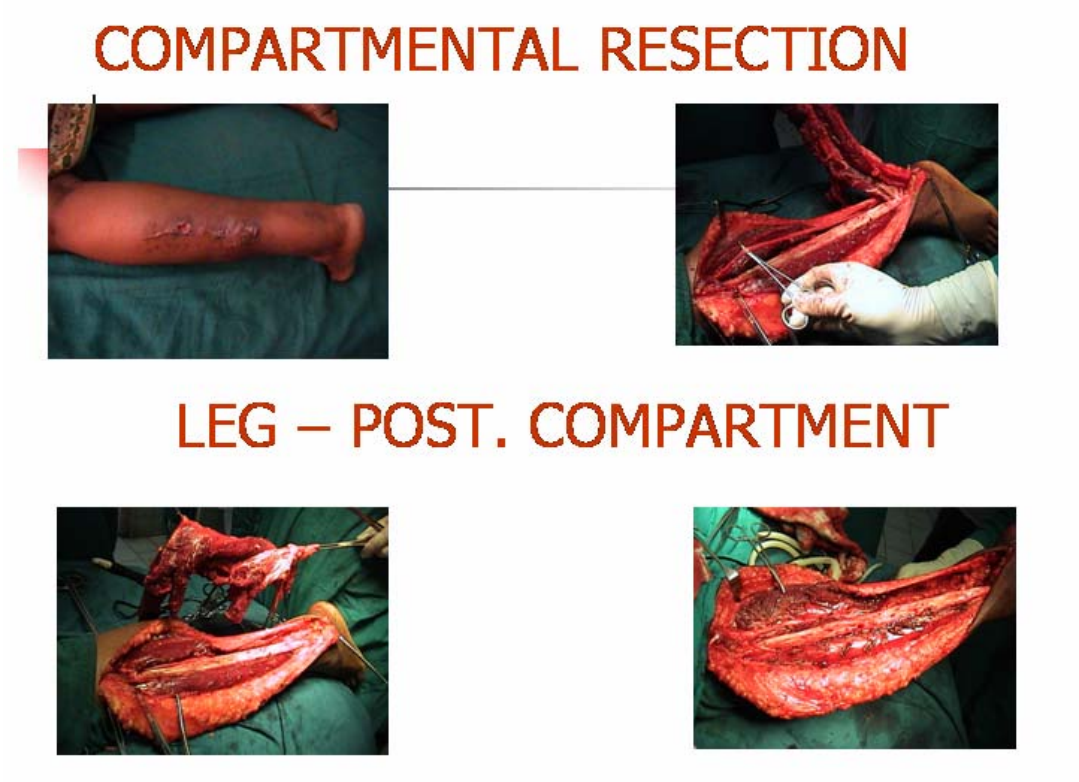
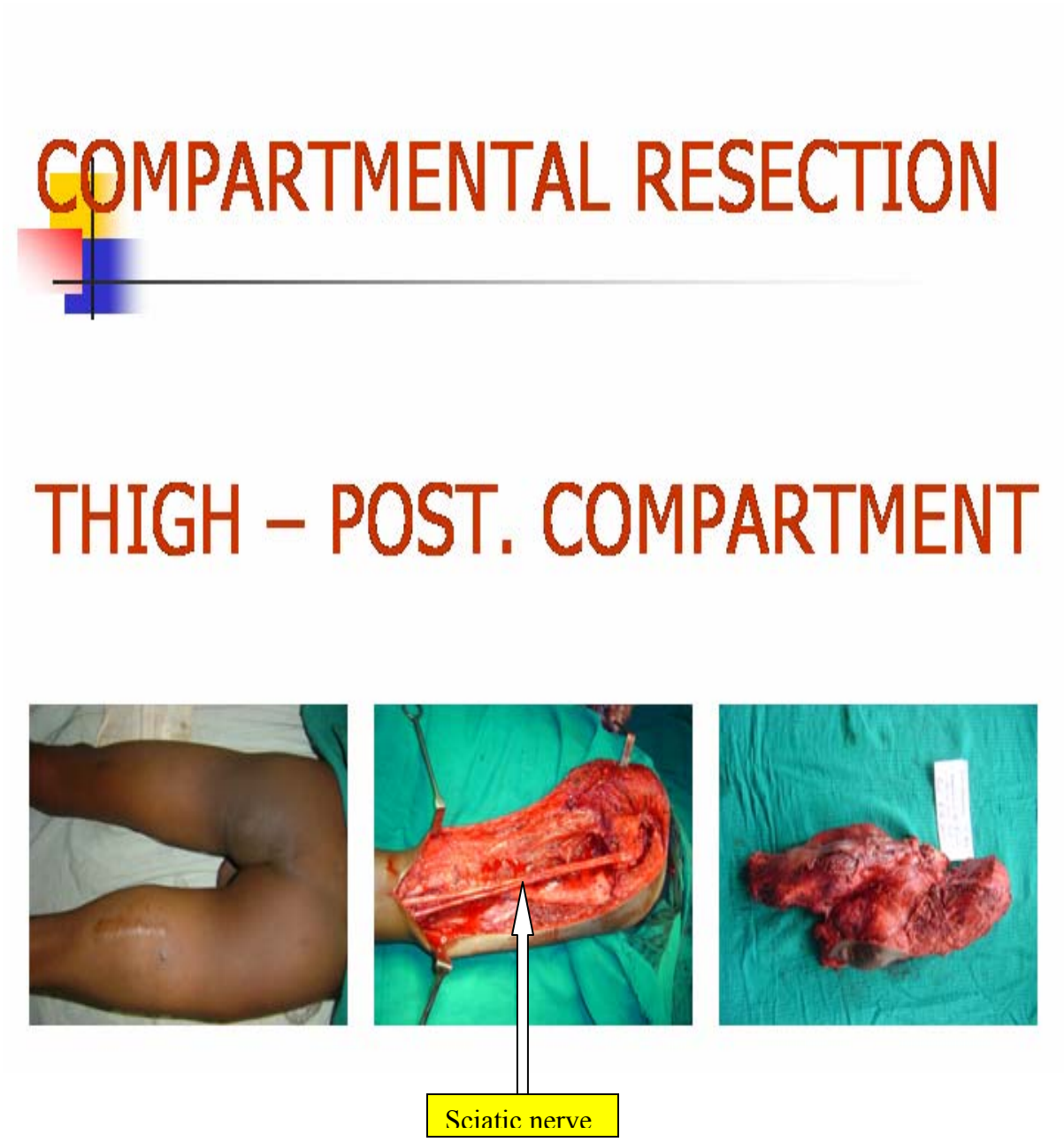
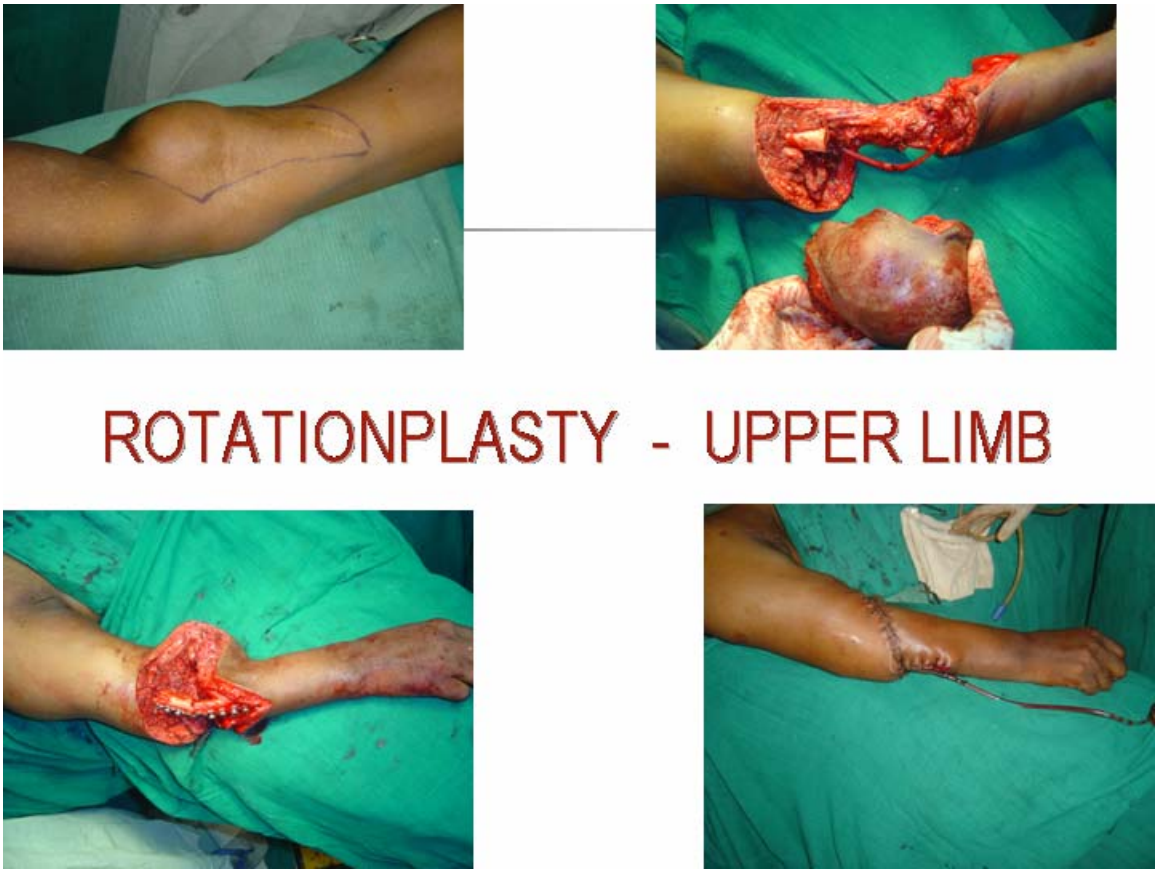


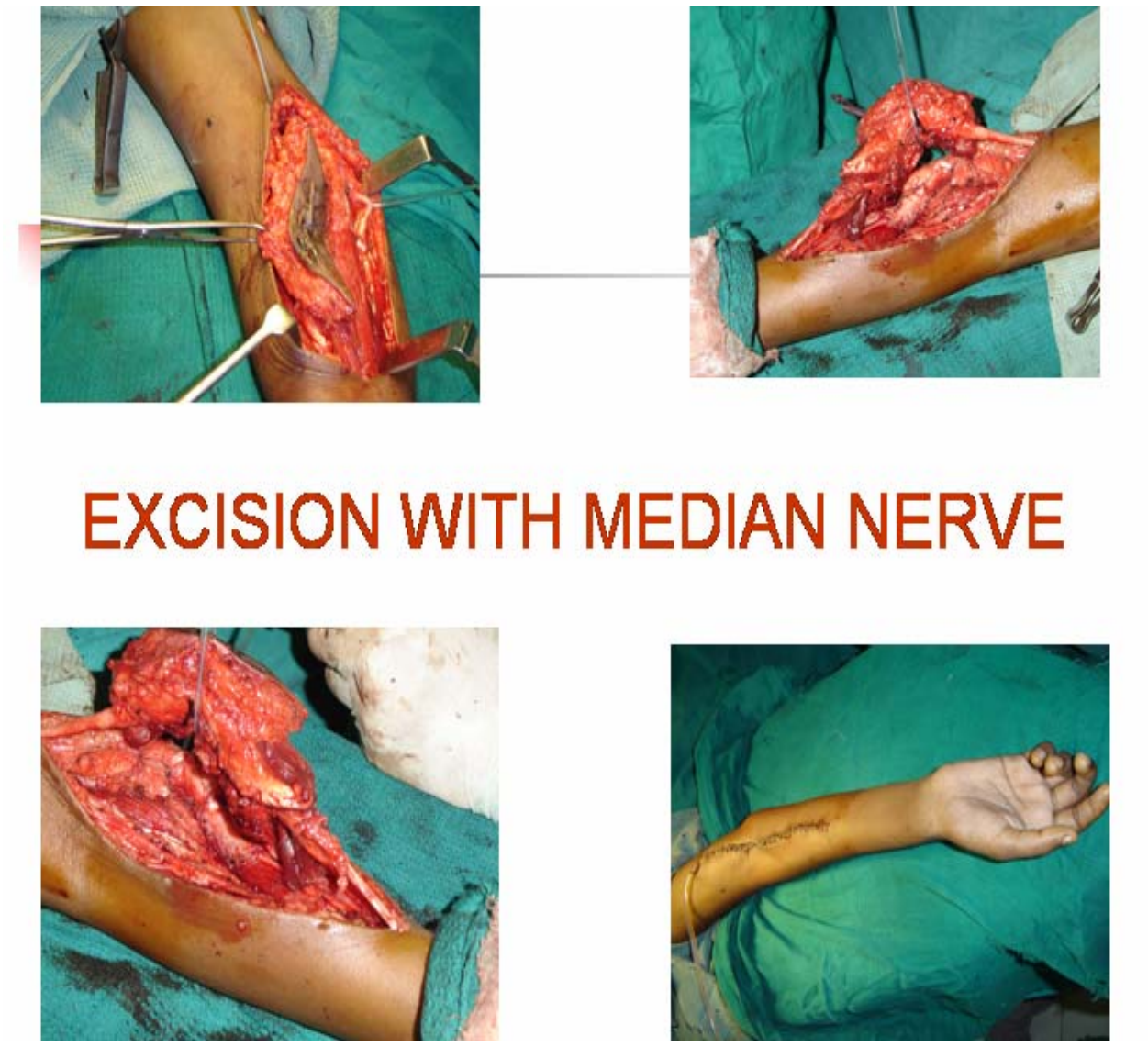
Figure illustrate, A 32 yrs old lady a case of MPNST (recurrence) Rt leg underwent post compartmental resection.



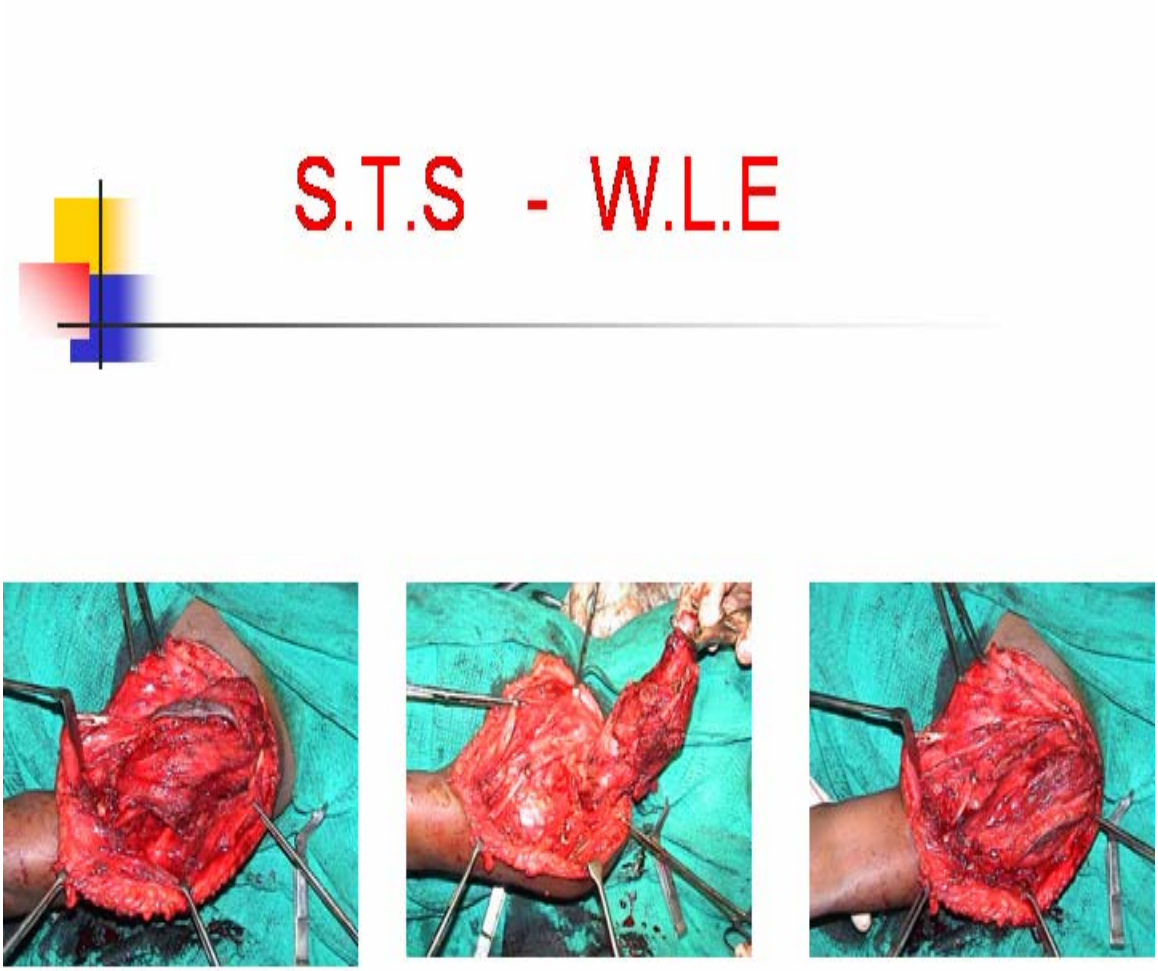
This figure illustrate, A 36 yrs old man , a case of MPNST Lt thigh underwent Hamstring compartmental resection.



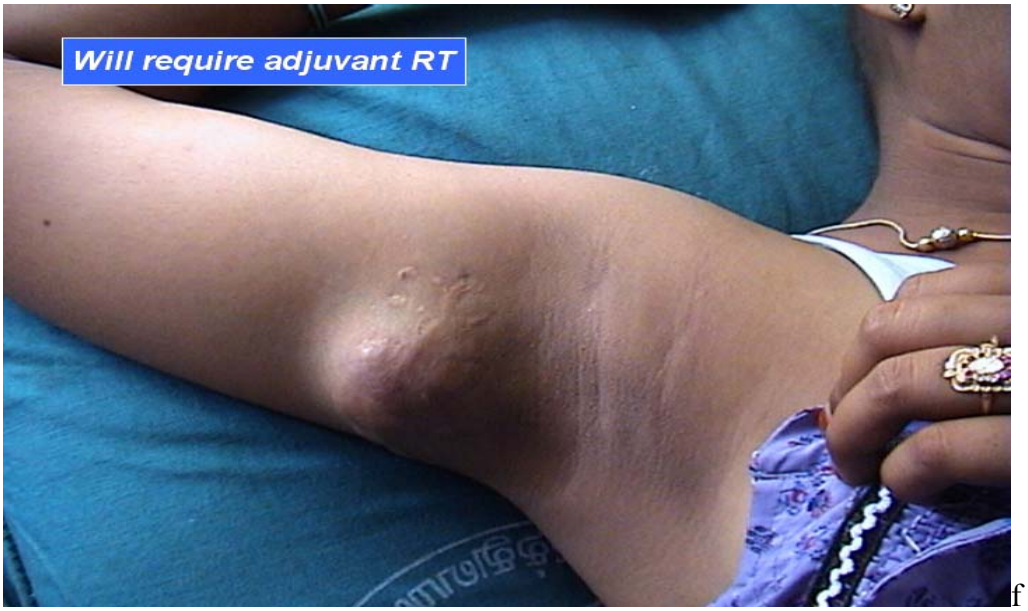
This figure illustrate, A 40yrs old lady , a case of Malignant fibrous histiocytoma Rt arm near elbow joint underwent AmputoResection and Rotationplasty.



This figure illustrate, A 34 yrs old lady , a case of MPNST Rt forearm , underwent Wide local excision with median nerve.



This figure illustrate A 36yrs old man , a case of Liposarcoma Lt thigh underwent Wide local excision.



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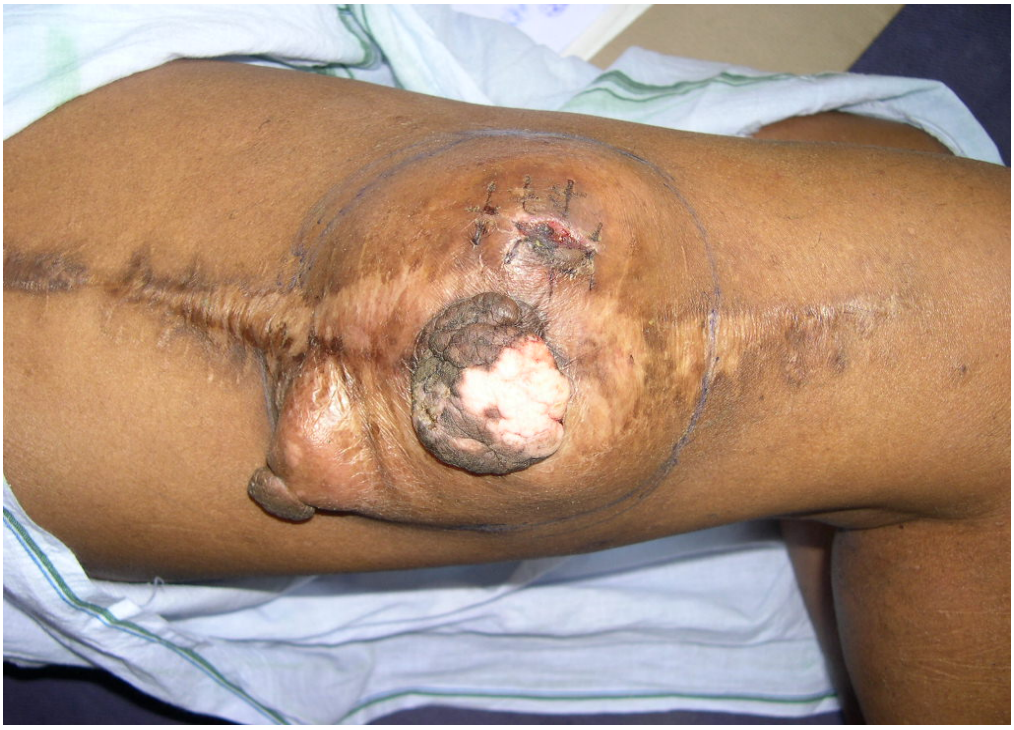
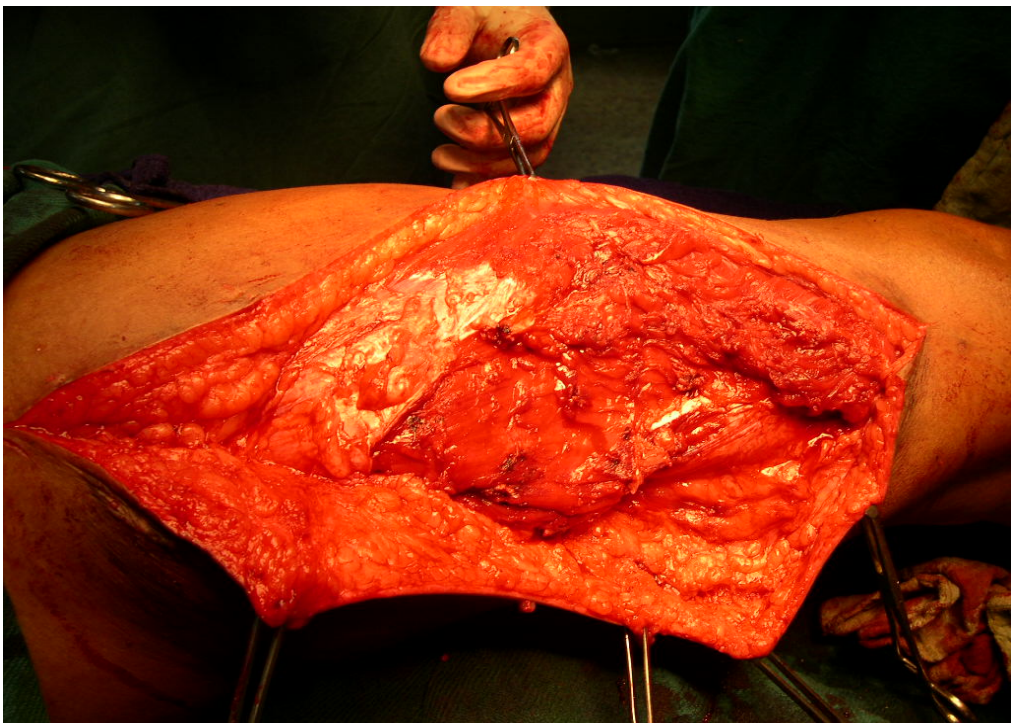
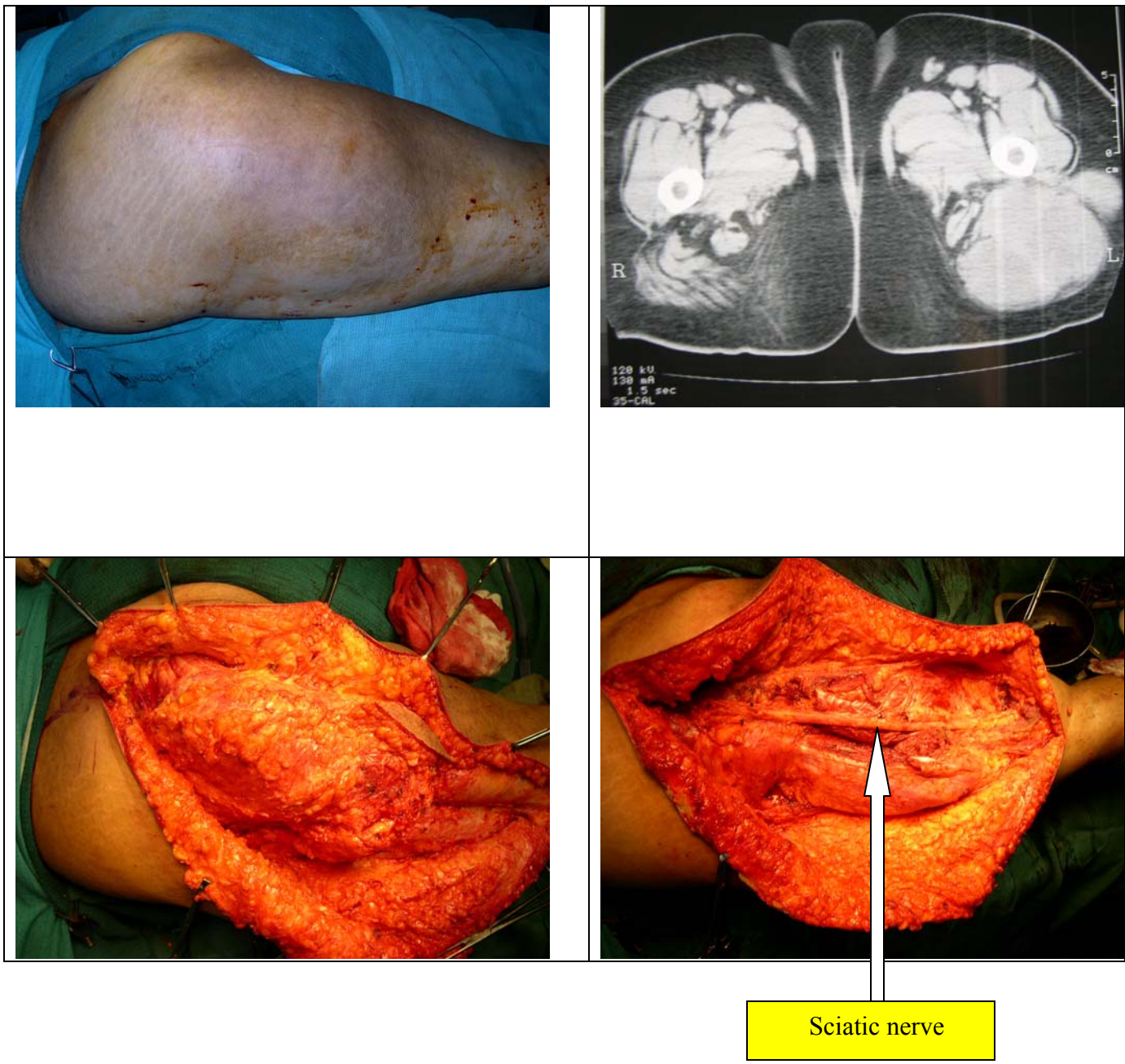
This figure illustrate, A 40yrs old lady , acase of High grade Liposarcoma Rt arm With local recurrence underwent wide local resection with post operative radiotherapy.

This figure illustrate A 55yrs old man a case of High grade malignt fibrous histiocytoma underwent multimodality treatment presented post radiotherapy complication..





This figures illustrate, A 45yrs old man ,a case of recurrent Malignant fibrous histiocytoma Rt thigh underwent Wide local excision .



This figure illustrate, A 56yrs male a case of Malignant fibrous histiocytoma Lt thigh posterior compartment, underwent Hamstring compartmental resection.