A Dissertation on

A STUDY ON RETROPERITONEAL SOFT TISSUE SARCOMA

Submitted to

The Tamilnadu Dr.M.G.R.Medical University

in partial fulfillment of the requirement

for the award of degree of

M.S. (GENERAL SURGERY)

BRANCH I

KILPAUK MEDICAL COLLEGE
THE TAMILNADU Dr.M.G.R.MEDICAL UNIVERSITY
CHENNAI, TAMILNADU

MARCH 2009

BONAFIDE CERTIFICATE

Certified that the dissertation titled **"A STUDY ON RETROPERITONEAL SOFT TISSUE SARCOMA"** is a bonafide work of the Candidate **Dr.RM.PALANIAPPAN,** carried under my supervision. Certified further that to the best of my knowledge the work reported herein does not form part of any other thesis or dissertation on the basis of which a degree or award was conferred on an earlier occasion on this or any other candidate.

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TO WHOMSOEVER IT MAY CONCERN

Dear Sir / Madam

Sub: General Surgery – MS PG's Dissertation Ethical Committee – Reg.
Ref: Requisition from H.O.D. General Surgery

This is in reference to the letter dated 2.1.2008 regarding Ethical committee meeting clearance with regard to the following topics

Sl.No	Name of the Post Graduate	Dissertation Topic
1	Dr.Valarmathi	Clinical study on Gastric outlet obstruction in 50 cases
2	Dr.RM.Palaniappan	A study on Retroperitoneal Sarcoma
3	Dr.Martin Paniraj	Study on Secondary neck nodes from squamous cell carcinoma
4	Dr.Krishna Kumar	A study on clinicopathological analysis of solitary nodule thyroid
5	Dr.Arul Kumar	Analytical study on ventral hernias presenting as surgical emergency
6	Dr.Emmanuel Thas	An analysis of obstructive jaundice
7	Dr.Skanda	A discussion of gut and genitourinary anomalies encountered in emergency surgery.
8	Dr.Sathish	Role of emergency gastrointestinal ostomies in present scenario
9	Dr.Madhu Sudhanan	Splenic abscess – an audit
10	Dr.Senthil Kumar	Abdominal tuberculosis
11	Dr.Suja	Profile of diagnosis and management of anorectal fistulae
12	Dr.Saravanan	Sigmoid volvulus presentation and management
13	Dr.ShivShankar	Analysis of scrotal swelling in Government Royapettah Hospital

We are glad to inform you that at the EC meeting held on 3.1.08 on the above topics were discussed and **Ethically approved**.

DEAN

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We confirm that no member of the study team is on the Ethics Committee and no member of the study team voted.

The trial will also follow the Ethics Guidelines for Bio-Medical Research On Human subjects issued by ICMR, New Delhi and will not involve any expense to the Government and will not be detrimental to the normal functioning of the Institution.

The study will also satisfy the revised order issued by the Government of Tamil Nadu, Health and Family Welfare Department G.O.MS.No:319, H & FW, Dept. dated 30.11.2001.

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CONTENTS

INTRODUCTION

A STUDY ON RETROPERITONEAL SOFT TISSUE SARCOMA

1. INTRODUCTION

Soft tissue sarcomas are the most frequent sarcomas. They are a rare and heterogeneous group of tumors that arise from the supporting extra skeletal tissues (i.e., muscle, fascia, nerve, connective, fibrous, and fatty tissues. Although soft tissues comprise 75% of the average body weight, these neoplasms represent less than 1% of all adult and 15% of pediatric malignancies. Soft tissue sarcomas are a disease of adulthood, occurring most commonly in persons between 30 and 60 years of age. The sole exception is rhabdomyosarcoma, which occurs in young children.

Each of the various soft tissue sarcomas has a unique morphology, biological behavior, and prognosis. However, like bone sarcomas, they all share certain biological and behavioral characteristics. The clinical, radiographic, and surgical management of most soft tissue sarcomas is identical, regardless of histogenesis.

The treatment of soft tissue sarcoma has become multidisciplinary, as advances in biology, imaging, surgery, chemotherapy and radiotherapy have improved the outlook for these patients who have these malignancies.

Fifteen percent of adult soft tissue sarcomas occur in the retroperitoneum. Most retroperitoneal tumors are malignant, and about one-third are soft tissue sarcomas. The most common sarcomas occurring in the retroperitoneum are liposarcomas, malignant fibrous histiocytomas, and leiomyosarcomas. In contrast to extremity sarcomas, local recurrence and intra-abdominal spread are frequent patterns of relapse for retroperitoneal tumors.

The size at presentation depends on the location. Tumors in the proximal extremities and retroperitoneum are often quite large, whereas distal extremity tumors are often small. The anatomic site of the primary disease represents an important variable that influences treatment and outcome. Soft tissue sarcomas of the extremities account for about 50% of all sarcomas, gastrointestinal (GI) sarcomas for 25%, retroperitoneal sarcomas for 15-20%, and head and neck for 9%. The most common subtypes of soft tissue sarcomas are malignant fibrous histiocytoma, liposarcoma, leiomyosarcoma, unclassified sarcoma, synovial sarcoma, and malignant peripheral

nerve sheath tumors; however, more than 50 different histologic subtypes of soft tissue sarcoma have been identified. Soft tissue sarcomas most commonly metastasize to the lungs; tumors arising in the abdominal cavity more commonly metastasize to the liver and peritoneum.

2. AIM OF STUDY

1. To study the incidence of retroperitoneal sarcoma in our institution.

2. To study age distribution and sex incidence.

3. To study stage of the disease at presentation.

4. To study the incidence of various pathological types.

5. To study the completion of resection of tumour and adjacent organs removed.

6. To study the incidence of the grade of the tumour.

3. REVIEW OF LITERATURE

Embryogenesis of Retroperitoneum

Normal Development

The peritoneum develops around the third week of embryonic life. Differentiation to mesothelial cells by the primitive mesodermal lining of the early fetal coelomic cavity produces the parietal and visceral layers.

The development of the retroperitoneal fasciae is enigmatic and obscure. The dorsal myotomes are responsible for the development of the psoas major and the quadratus lumborum muscles. The ventral myotomes are responsible for the genesis of the transversus abdominis muscle. Perhaps both myotomes are responsible for the genesis of these periparietic fasciae, which are united at the lateral border of the psoas major muscle.

The transversalis fascia and other fasciae related to the lumbar musculature are of mesodermal origin. The muscles of the trunk are derived from dorsal myotomes of truncal somites and characteristically maintain their innervation from the segmental spinal nerves at the levels of the origin of the muscles.

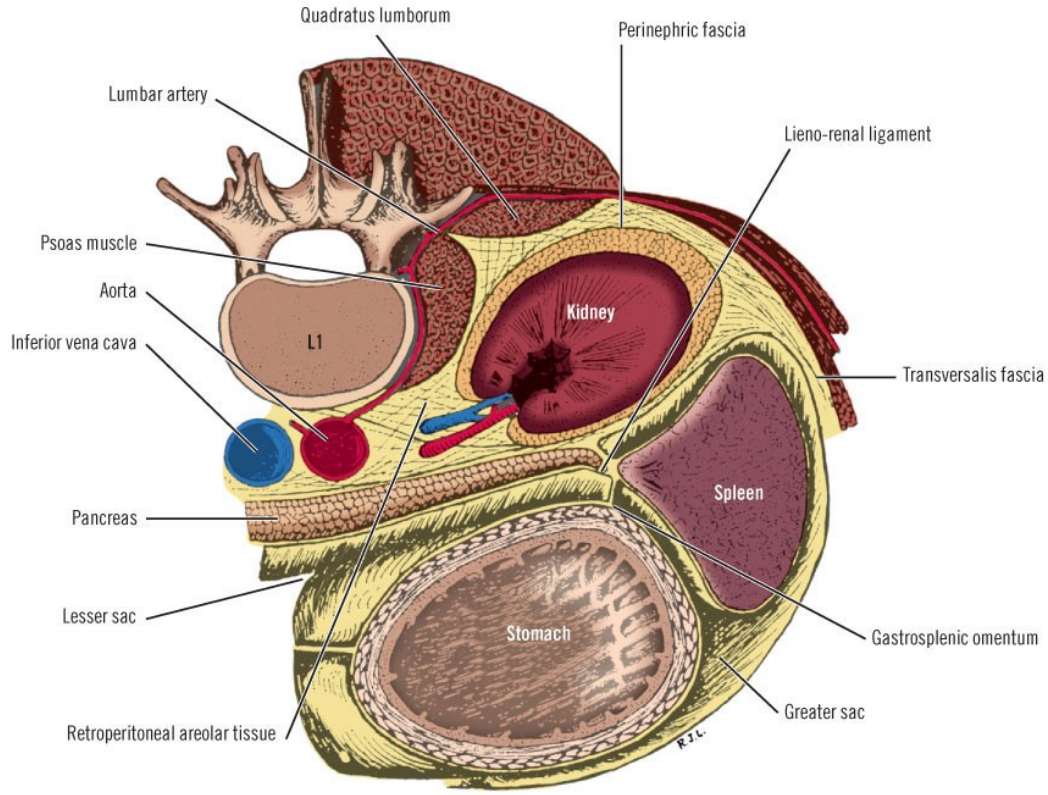
Surgical Anatomy

The retroperitoneal space is the area of the posterior abdominal wall that is located between the parietal peritoneum and the deep or internal surface of the transversalis fascia. Within this space are embryologically related organs which are referred to as the retroperitoneal viscera. These include the adrenals, kidneys, and ureters. There are also numerous vascular and neural structures, including the aorta and its branches, the inferior vena cava and its tributaries, the lymphatics and the lymph nodes, the lumbar plexus with its branches, and the sympathetic trunks.

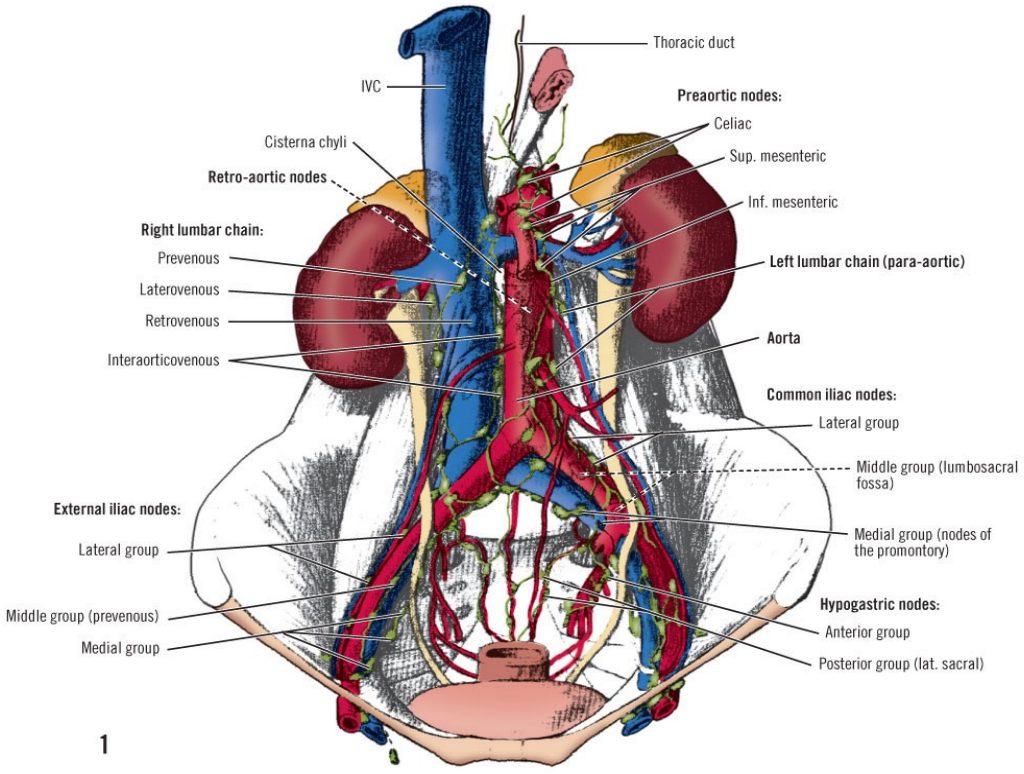
In addition to the organs and tissues that develop in the retroperitoneum, several other organs attain a secondarily retroperitoneal position in later embryologic development. These include most of the duodenum, the pancreas, and major portions of the ascending and descending colon.

Within the greater retroperitoneal space, there are also several small spaces, or subcompartments. Loose connective tissue and fat surround the anatomic entities, and, to a variable degree, occupy the smaller spaces. The parietal peritoneum is in continuity with the visceral peritoneum, and vice versa.

Figure



showing the important structures in retroperitoneum



Compartments of the Retroperitoneal Space

Three compartments of the retroperitoneal space are related to the kidney:

1.Anterior pararenal compartment

2.Posterior pararenal compartment

3.Perirenal compartment

The renal fascia, a collagenous connective tissue of mesodermal origin enveloping the kidney, is responsible for this compartmentalization.

The fascial layers and the spaces related to the kidney are as follows, from anterior to posterior:

- Peritoneum
- Anterior pararenal space (with a variable quantity of loose connective tissue and fat)
- Anterior lamina of Gerota's fascia
- Perirenal space (the kidney and the ureter; the adrenal in a separate subcompartment; fat)
- Posterior lamina of Gerota's fascia

- Posterior pararenal space (usually with a large content of more compact fat)

- Thoracolumbar (lumbodorsal) fascia and the fascia of the psoas muscle

To generalize, the muscle fascia lining the abdomen is referred to as the transversalis fascia. More specifically, however, the transversalis fascia, which is the fascial lining of the transversus abdominis muscle, is continuous with the subdiaphragmatic fascia above. Medially, it is continuous with the psoas fascia and the thoracolumbar (or lumbodorsal) fascial investment (anterior lamina) of the quadratus lumborum muscle. Below, it is continuous with the fascia of the iliacus muscle and the parietal muscular fascia of the true pelvis.

Retroperitoneal Lymphatics

From an anatomic standpoint, the retroperitoneal lymph nodes can be rather difficult to classify. The retroperitoneal lymphatics form a very rich and extensive chain from the inguinal ligament and pelvis to the respiratory diaphragm and posterior mediastinal nodes. Usually, these lymph nodes are located close to the aorta and IVC. The right paraaortic lymph nodes are in very close relationship with the left paracaval lymph nodes. The number of abdominal and pelvic lymph nodes is approximately 230.

The following classification is very simple, logical, and anatomic:

- Aortic Group

– Preaortic nodes

– Retroaortic nodes

– Paraaortic nodes

- Caval Group

– Precaval (prevenous) nodes

– Retrocaval (retrovenous) nodes

– Paracaval (laterovenous) nodes

- Pelvic Group

– Common iliac nodes

– External iliac nodes

– Internal iliac (hypogastric) nodes

– Obturator nodes

– Sacral nodes

Retroperitoneal Innervation

Six nerves and the lumbar sympathetic chains are present in the retroperitoneal space. The six nerves are branches of the lumbar plexus, which is formed by a branch of T12 as well as by the anterior primary rami of the first four lumbar nerves. Most of the branches of the plexus are related to the psoas major muscle, passing through it or behind it or being formed within it.

The nerves formed by the plexus are:

- Iliohypogastric
- Ilioinguinal
- Genitofemoral
- Lateral femoral cutaneous
- Obturator
- Femoral

The retroperitoneum can be approached and explored by several routes, including the transperitoneal route and the extraperitoneal route. There are two accepted procedures used for diagnosis of retroperitoneal injuries and for exploration of clinicopathological entities. The Cattell maneuver exposes right-sided structures. The Mattox maneuver exposes left-sided structures.

Cattell maneuver

Step 1. Incise the lateral peritoneum along the cecum, ascending colon, and hepatic flexure

Step 2. Divide the white line of Toldt (peritoneal reflection at the area of the lateral wall of the cecum and ascending colon)

Step 3. Perform duodenal mobilization (Kocherization)

Step 4. Mobilize all right-sided anatomic entities anteromedially

Mattox maneuver

Step 1. Incise the lateral peritoneum along the splenic flexure, descending colon, and upper sigmoid

Step 2. Divide the white line of Toldt

Step 3. Carefully mobilize the spleen, including the pancreatic tail, stomach, and left colon

Step 4. Gently push all left-sided anatomic entities anteromedially

History

The word sarcoma dates to Galen and the Greek term describing a fleshy growth. The idea of a sarcoma as a distinct type of cancer was not formalized until the mid 1800s by Virchow.

Incidence

Soft tissue sarcoma represents less than 1% of all adult and 15% of pediatric malignancies. Retroperitoneal sarcomas are rare tumors accounting for only 1%–2% of all solid malignancies. Of all sarcomas, the majority occur outside of the retroperitoneum. Only 10%–20% of sarcomas are retroperitoneal sarcomas, and the overall incidence is 0.3%–0.4% per 100000 of the population . The peak incidence is in the 5th decade of life, although they can occur in any age group.

Distribution

Soft tissue sarcoma can occur in any site throughout the body. Forty three percent are in extremities with two thirds of extremity lesions occurring in the lower limb, and 34% are intra-abdominal, divided between visceral (19%) and

retroperitoneal (15%) lesions. Trunk sarcoma occurs in 10% of individuals and others in 10% of patients.

Etiology

Most soft tissue sarcomas have no clearly defined cause, although multiple associated or predisposing factors have been identified.

Various genetic syndrome which are predisposing to soft tissue sarcoma are neurofibromatosis type 1, retinoblastoma, LI-Fraumeni syndrome, Gardner's syndrome, Werner's syndrome, Goblin's syndrome, Carney's triad and tuberous sclerosis.

Radiation therapy is the known cause of soft tissue sarcoma. They are most often seen in diseases that are commonly treated with radiotherapy and in those in which a long survival period is expected. The prime candidate diseases are breast cancer, lymphoma, and cervical cancer. The children are at risk due to time latency involved. It arises close to penumbra of radiotherapy fields.

Classification

It is based on line of differentiation i.e. the type of tissue formed rather than from the type of origin. WHO's classification is used widely.

- Fibrous tumors
- Fibrohistiocytic tumors
- Lipomatous tumors
- Smooth muscle tumors
- Skeletal muscle tumors
- Tumors of Blood vessels & lymphatics
- Perivascular tumors
- Synovial tumors
- Mesothelial tumors
- Peripheral N. sheath tumors
- Primitive neuroectodermal tumors(PNET)
- Extra skeletal osseous & cartilagenous tumors
- Miscellaneous tumors

The basis cell appearance on smears, they are classified clinically as follows

- Myxoid tumors
- Spindle cell tumors
- Pleomorphic tumors
- Polygonal tumors
- Round cell tumors
- Miscellaneous

Pathology

The most common types of retroperitoneal soft tissue sarcomas in adults vary from study to study. However, in most studies, the most frequently encountered cell types are liposarcomas, leiomyosarcomas and malignant fibrous histiocytomas (MFH) . Recently, the frequent diagnosis of MFH in the retroperitoneum has been-disputed. With the use of immunohistochemistry, many of these fibrous tumors have now been shown to represent other sarcoma types such as leiomyosarcomas or dedifferentiated liposarcomas

Retroperitoneal sarcomas generally present as large masses; nearly 50% are larger than 20 cm at the time of diagnosis. They typically do not produce symptoms until they grow large enough to compress or invade contiguous structures. The differential diagnosis of a retroperitoneal tumor includes lymphoma, germ-cell tumors, and undifferentiated carcinomas. The overall prognosis for patients with retroperitoneal tumors is worse than that for patients with extremity sarcomas.

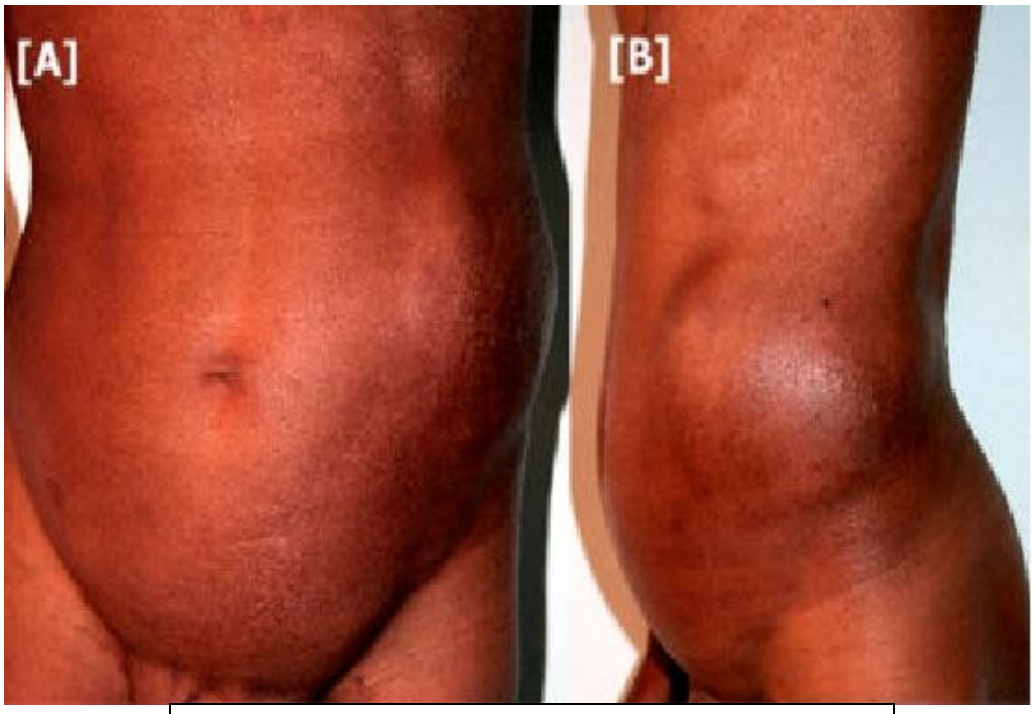


Fig 1 – showing retroperitoneal tumor presenting as abdominal mass (A) AP view ; (B) Lateral view

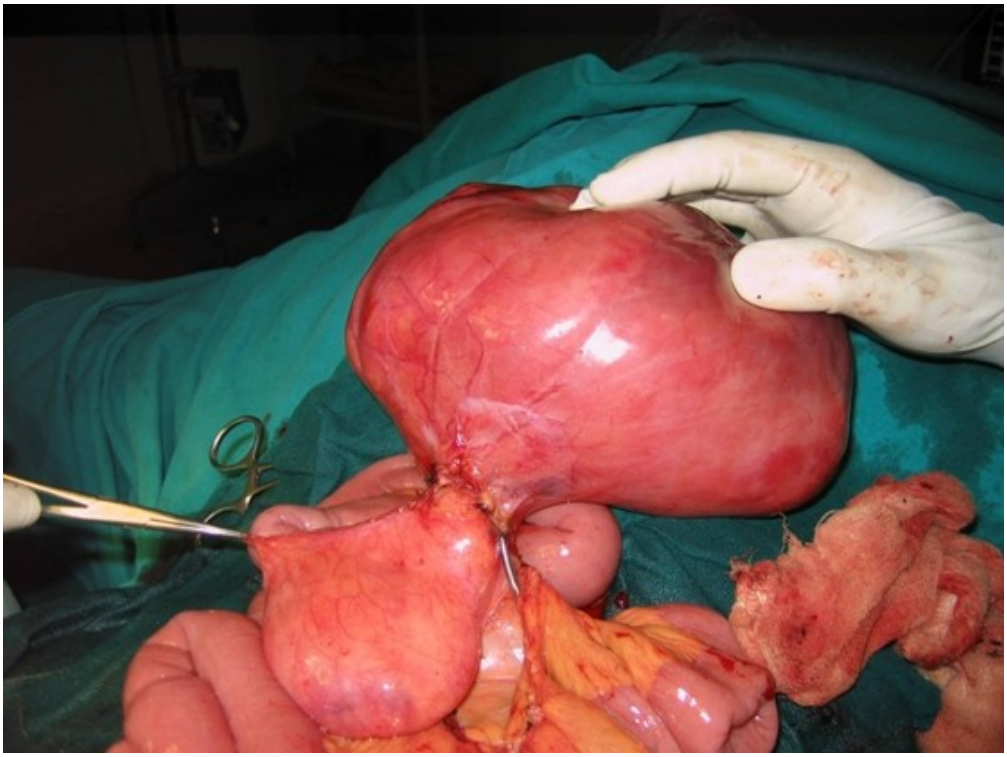


Fig 2 – Intra-operative picture of retroperitoneal sarcoma seen involving adjacent bowel loops

Grading of sarcoma

After establishing the diagnosis of sarcoma, the most critical piece of information the pathologist can provide to the clinician is histologic grade. This remains the most important prognostic factor for determining disease-free and overall survival rate.

The pathologic features that define grade include cellularity, differentiation, pleomorphism, necrosis, and number of mitoses.

Unfortunately, the criteria for grading are neither specific nor standardized. Several grading scales and systems are used: a four-grade system (Broder's), a three-grade system (low, intermediate, high) such as National Cancer Institute (NCI) grading system and that of the French Federation of Cancer Centers Sarcoma group, and a binary system (low, high) as is used at Memorial Hospital.

Many pathologists consider mitotic activity and degree of necrosis to be the most important pathologic features. To define a practical grading system, the European Organization for Research and treatment of cancer (EORTC) conducted a study in which, the multivariate analysis showed only mitotic count (fewer than

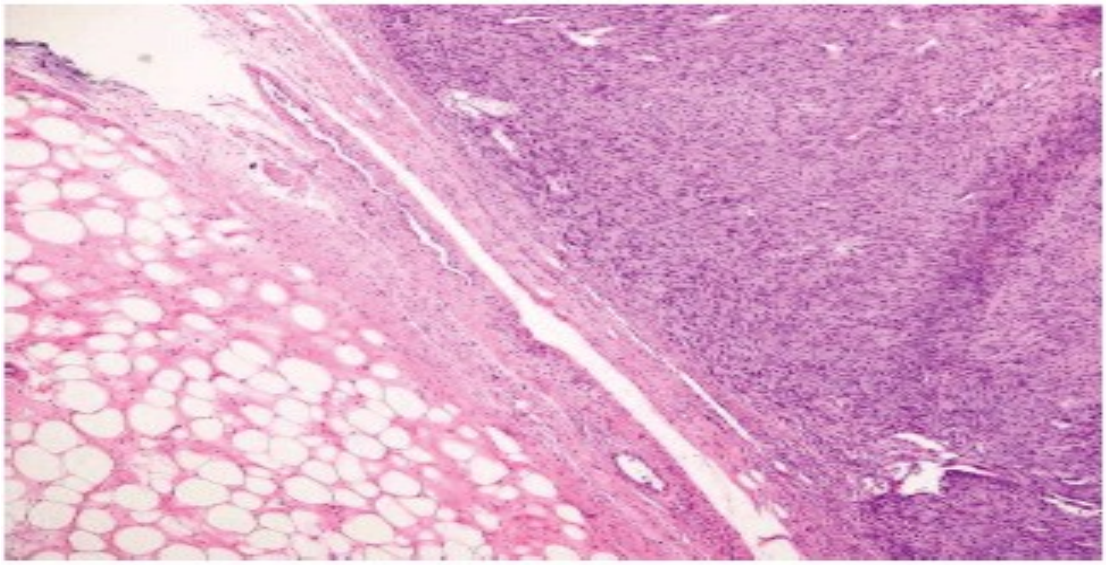


Figure 3 – Photomicrograph showing the border between the two histopathological variants: to the left, the well-differentiated liposarcoma; and, to the right, the dedifferentiated liposarcoma (hematoxylin and eosin staining, magnification of ×100).

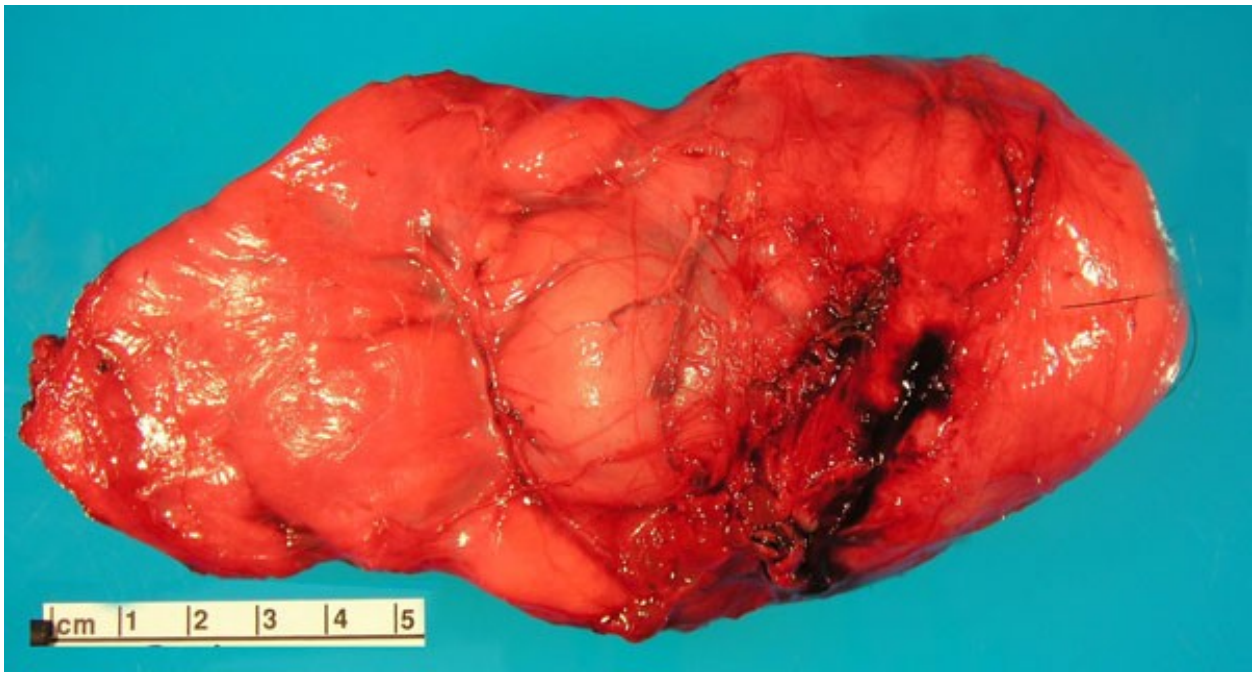


Fig 4- showing a specimen of retroperitoneal liposarcoma

3, 3 to 20, and more than 20 mitoses per 10 consecutive high-power fields), the presence or absence of necrosis, and tumor size predicted survival.

Several tumors that are considered sarcomas have no recognizable normal tissue counterpart (e.g., alveolar soft part tumor, Ewing's sarcoma, Epithelioid sarcoma). These tumors often have unique clinical features and usually are not graded. In 2002 AJCC/TNM staging system of sarcoma, only two grades, low versus high are used to stage soft tissue sarcomas. To accurately determine tumor grade, an adequate tissue sample must be well fixed, well stained, and reviewed by an experienced sarcoma pathologist.

Staging

Staging has an important role in determining the most effective treatment of soft tissue sarcomas. The stage is determined by the size of the tumor, the histologic grade, and whether it has spread to lymph nodes or distant sites. For complete staging, a thorough physical examination, x-rays, laboratory studies, and careful review of all biopsy specimens (including those from the primary tumor, lymph nodes, or other suspicious lesions) are essential. CT chest done to find out lung metastases. CT abdomen and pelvis to find out the extension of tumour and liver metastases.

The staging system applies to all soft tissue sarcomas except Kaposi's sarcoma,

dermatofibrosarcoma, infantile fibrosarcoma, and angiosarcoma. In addition, sarcomas arising within the confines of the dura mater, including the brain, and sarcomas arising in parenchymatous organs and from hollow viscera are not optimally staged by this system.

Data to support this staging system are based on current analyses from multiple institutions and represent the recommendations of an AJCC task force on soft tissue sarcoma. In the era of cytoreductive neoadjuvant treatments, clinical and pathologic staging may be altered in the future. Because pathologic staging drives adjuvant therapy decisions, patients should be restaged after neoadjuvant therapies have been administered.

Histologic type, grade, and tumor size and depth are essential for staging. Histologic grade of sarcoma is one of the most important parameters of the staging system. Grade is based on analysis of various pathologic features of a tumor, such as histologic subtype, degree of differentiation, mitotic activity, and necrosis. Accurate grading requires an adequate sample of well-fixed tissue for evaluation.

Accurate grading is not always possible on the basis of needle biopsies or in tumors that have been previously irradiated or treated with chemotherapy.

Inclusions

The present staging system applies to soft tissue sarcomas. Primary sarcomas can arise from a variety of soft tissues. These tissues include fibrous connective tissue, fat, smooth or striated muscle, vascular tissue, peripheral neural tissue, and visceral tissue.

Regional Lymph Nodes

Involvement of regional lymph nodes by soft tissue sarcomas is uncommon in adults. When present, regional nodal disease has prognostic significance similar to that of visceral metastatic disease.

Metastatic Sites

Metastatic sites for soft tissue sarcoma are often dependent on the original site of the primary lesion. For example, the most common site of metastatic disease for patients with extremity sarcomas is the lung, whereas retroperitoneal and gastrointestinal sarcomas often have liver as the first site of metastasis.

TNM / The American Joint Committee on Cancer (AJCC) has designated staging by the four criteria of tumor size, nodal status, grade, and metastasis (TNGM).

Grade and TNM Definitions

Tumor grade (G)

- GX: Grade cannot be assessed
- G1: Well differentiated
- G2: Moderately differentiated
- G3: Poorly differentiated
- G4: Poorly differentiated or undifferentiated

Primary tumor (T)

- TX: Primary tumor cannot be assessed
- T0: No evidence of primary tumor
- T1: Tumor 5 cm or less in greatest dimension
    - T1a: Superficial tumor
    - T1b: Deep tumor
- T2: Tumor 5 cm or larger in greatest dimension
    - T2a: Superficial tumor

- T2b: Deep tumor

Regional lymph nodes (N)

- NX: Regional lymph nodes cannot be assessed
- N0: No regional lymph node metastasis
- N1: Regional lymph node metastasis [Note: Presence of positive nodes (N1) is considered stage IV.]

Distant metastasis (M)

- MX: Distant metastasis cannot be assessed
- M0: No distant metastasis
- M1: Distant metastasis

NOTES

1. Superficial tumor is located exclusively above the superficial fascia without invasion

of the fascia; deep tumor is located either exclusively beneath the superficial fascia, superficial to the fascia with invasion of or through the fascia, or both superficial yet beneath the fascia. Retroperitoneal, mediastinal, and pelvic sarcomas are classified as deep tumors.

2. Ewing's sarcoma is classified as G4.

STAGE GROUPING

IA	T1a	N0	NX	M0	G1–2	G1	Low
	T1b	N0	NX	M0	G1–2	G1	Low
IB	T2a	N0	NX	M0	G1–2	G1	Low
	T2b	N0	NX	M0	G1–2	G1	Low
IIA	T1a	N0	NX	M0	G3–4	G2–3	High
	T1b	N0	NX	M0	G3–4	G2–3	High
IIB	T2a	N0	NX	M0	G3–4	G2–3	High
III	T2b	N0	NX	M0	G3–4	G2–3	High
IV	Any T	N1		M0	Any G	Any G	High or Low
	Any T	Any N		M1	Any G	Any G	High or Low

Evaluation and Workup

The initial evaluation and workup for retroperitoneal abdominal soft tissue sarcomas are similar to that for the extremity sarcomas. This workup involves a thorough H&P and appropriate imaging studies, including an abdominal and pelvic CT with contrast with or without an MRI. Chest imaging with a plain radiograph or CT should be done, especially for patients whose tumors warrant preoperative or

postoperative chemotherapy. If possible, the patient should be reviewed by a multidisciplinary sarcoma panel. Note that for staging, all retroperitoneal lesions are considered to be deep lesions. MRI is preferred for extremity sarcomas, whereas CT is preferred for retroperitoneal sarcomas.

Radiologic assessment should include CT of the abdomen and pelvis to define the extent of the tumor and its relationship to surrounding structures, particularly vascular structures.Imaging should also encompass the liver for the presence of metastases, the abdomen for discontiguous disease, and the kidneys bilaterally for function. Thoracic CT is indicated to detect lung metastases. For patients presenting with an equivocal history, an unusual appearance of the mass, an unresectable tumor, or distant metastasis, CT-guided core-needle or laparoscopic biopsy is appropriate to obtain a sample for tissue diagnosis

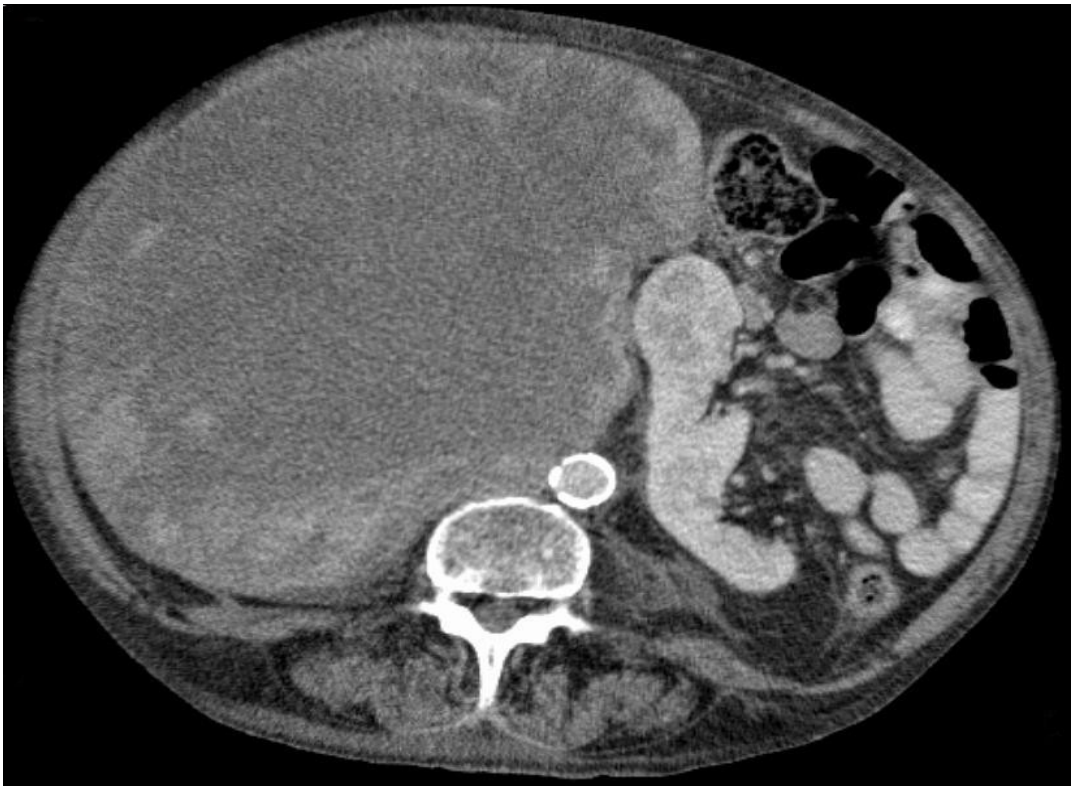


Fig 5 – Contrast enhanced CT Abdomen showing a huge retroperitoneal liposarcoma pushing the bowel loops to opposite side

18Fluorodeoxyglucose-positron emission tomography (18FDG-PET) scan may be useful for prognostication, grading and to assess response to chemotherapy. Tumor metabolism data acquired by FDG-PET will be useful in accurate grading and prognostication in sarcoma. Recent reports in literature have demonstrated the value of FDG-PET scan in evaluating response to neoadjuvant chemotherapy in patients with high-grade extremity soft tissue sarcomas, prediction of outcome in liposarcoma.

BIOPSY

The differential diagnosis of retroperitoneal abdominal soft tissue mass includes malignant lesions (such as other sarcomas, GIST, lymphomas, or germ cell tumors), desmoids, and benign lesions. The need for a biopsy remains somewhat controversial, and this decision should be based on the clinician's degree of suspicion that another malignancy is possible. Proof of the histologic subtype by biopsy is necessary for patients before receiving preoperative chemotherapy or RT; a CT-guided core biopsy is preferred. The goal of this strategy is to avoid inappropriate major resection of another tumor, such as an intra-abdominal lymphoma or germ cell tumor. If a retroperitoneal sarcoma is encountered unexpectedly at the time of laparotomy performed for some other reason, a core biopsy should be done to

establish the diagnosis as well as the histopathologic type and grade of tumor. Then, the optimal subsequent resection could be performed.

Value of Trucut Biopsy

In general, the important issue is the adequacy of the sample. Sufficient viable tissue is required that is both representative of the lesion and available for histopathologic evaluation, immunohistochemistry, and, when necessary, electron microscopy. As molecular markers become a factor in diagnosis, meticulous attention to the adequacy of biopsy, tissue preservation, and evaluation will be paramount.

Histopathologic interpretation varies from center to center and may be a major variable in decision making. As with other relatively rare lesions, it is essential that review of the histopathologic findings be made by an experienced group. More recent studies show improved diagnostic accuracy and confluence of opinion, at least for malignancy and grade.

Fine-Needle Aspiration Cytology

Fine-needle aspiration (FNA) cytology has been examined by a number of authors. Some authors have argued that biopsy itself is not justified if FNA is available. But it is usually confined to the confirmation of recurrence rather than used for the primary diagnosis.

The use of FNA in patients with large sarcomas who are candidates for neoadjuvant therapy to improve survival is also problematic due to difficulty in grading and subtyping these tumors accurately from such small samples.

Frozen Section

In some institutions, frozen section is relied on as the diagnostic tool of choice. For diagnosis of malignancy, frozen section is accurate, but for histopathologic subtypes and grade, it is inferior to permanent sections of either Trucut or incisional biopsy.

Frozen section can guide retrieval of adequate diagnostic material and, depending on the initial evaluation, can be an important triage mechanism to direct further pathologic workup. However, open biopsy with the help of frozen-sectioning support may be indicated when the Trucut biopsy result is equivocal or for other clinical reasons.

Fatty lesions are not suitable for frozen-section evaluation, because of a loss of diagnostic material during frozen sectioning and other technical difficulties. In addition, freezing compromises the final interpretation on permanent sections.

Important application of frozen section is assessment of margin of resection. Negative margins of resection can be obtained by using this technique intraoperatively.

Immunohistochemistry

As an ancillary technique, immunohistochemistry is an invaluable tool that provides excellent information in assisting the surgical pathologist in establishing a precise diagnosis, as well as providing relevant prognostic and therapeutic information.

One of its major utilities is to correctly identify a tumor as being of mesenchymal or nonmesenchymal origin. Once mesenchymal origin has been established, histologic subtyping according to specific cell lineage may be achieved with the use of lineage-specific markers. Tumors of uncertain cell lineage and tumors with primitive small round cell morphology are often characterized by a unique immunohistochemical phenotype. In this group of tumors, immunohistochemistry is most widely applied and is of greatest value. By diagnosing the small round cell tumors with aid of immunohistochemistry, line of management is differed from spindle cell soft tissue tumors.

Despite the rapid development of molecular genetic techniques, immunohistochemistry still remains the most important diagnostic tool in the diagnosis of soft tissue tumors aside from recognition of morphologic features and clinical correlation.

Primary Treatment

Surgery is the standard treatment for retroperitoneal abdominal sarcomas. Complete surgical resection or macroscopic surgical resection is only achieved in less than 70% of patients with primary retroperitoneal sarcomas, because they often are near vital structures. Local recurrence occurs in approximately half of the patients who have undergone complete resection. Multimodality treatment is usually favored for retroperitoneal sarcomas due to the inability to obtain negative margin resections and high local recurrence rates. Preoperative RT is often preferred, because the volume of abdominal organs in the RT fields is smaller and it may render unresectable tumors more amenable to resection. Preoperative chemotherapy may have advantages over postoperative chemotherapy. However, the role of adjuvant RT or preoperative chemotherapy vs. postoperative chemotherapy has not yet been evaluated in randomized clinical trials.

Primary treatment depends on the resectability of the sarcoma. Biopsy is performed only if preoperative therapy is considered. CT-guided core biopsy is preferred.

Once the diagnosis is made, the surgical team needs to determine if the retroperitoneal sarcoma can be resected. Therefore one of the first determinations to be made is whether the tumor is localized, its local extent, and also if there is evidence of intra- or extra-abdominal metastatic spread of tumor. The location and size of the tumor, its relationship to adjacent organs, presence or absence of local extension, relationship to and/or involvement of major vascular structures, as well as the presence of normal anatomic variants and anomalies of major abdominal arteries and veins, are all crucial pieces of information that need to be provided. Since resection of one kidney is not uncommon, any radiographic evidence of unilateral renal dysfunction involving the kidney that is not adjacent to the tumor should be relayed to the surgical team. While it may be unavoidable that the patient will be left with a single poorly functioning kidney, the surgeon must be provided with all relevant information prior to attempted tumor resection.

Preoperative RT or preoperative chemotherapy (for chemo sensitive histologies) could be considered. Although most patients with retroperitoneal sarcomas (which are often liposarcomas) could be managed with surgical resection with or without intraoperative RT (IORT), the options for other therapy should be discussed, especially if incomplete resection is a reasonable probability. Long-term results of

two prospective trials showed favorable 5-year local recurrence-free (60%), disease-free (46%) and overall survival rates (61%) among patients who had R0 or R1 resection after preoperative RT for intermediate or high grade retroperitoneal sarcoma.86 Postoperative RT could be considered in patients with pathologic findings of high grade disease following negative margin resection or for microscopic positive margins (R1 resection). Macroscopic positive margins (R2 resection) should be managed as unresectable disease. Unresectable retroperitoneal sarcomas are defined as tumors that involve unresectable vital structures or tumors whose removal would cause unacceptable morbidity. Biopsy is recommended before any treatment for a patient with unresectable or metastatic retroperitoneal sarcoma . Patients with unresectable or metastatic disease have several options for primary treatment after biopsy including chemotherapy or RT to downstage tumors prior to resection. In asymptomatic patients, palliative surgery for symptom control, best supportive care, or observation are additional options. Unresectable tumors that become resectable following primary chemotherapy or RT should be managed as described under resectable disease.

Following primary treatment, if patients have progressive disease or remain unresectable with no downstaging of tumor, management decisions depend on whether patients are symptomatic or asymptomatic. Observation is considered for asymptomatic patients, whereas for symptomatic patients, treatment options are similar to those listed under primary treatment for unresectable or metastases.

Guidelines for Radiation Therapy

Sophisticated treatments with intensity-modulated radiation therapy (IMRT) and proton-beam should be considered to improve therapeutic effect.If resections with microscopically positive or grossly positive margins are anticipated, surgical clips should be left in place to identify high risk areas for recurrence, particularly for retroperitoneal or intra-abdominal sarcomas . Total doses of RT should be determined by normal tissue tolerance.

Preoperative RT

The usual dose of preoperative RT is 50 Gy. An intraoperative boost or a postoperative boost with brachytherapy or an external-beam RT is recommended for positive or close margins. Preoperative RT has several advantages. First, the treatment volume is smaller, because the need to cover the operative field is not present. Second, preoperative

radiation may reduce seeding during surgical manipulation of the tumor. The tumor may or may not regress with preoperative RT, but the pseudocapsule may thicken and become acellular, easing resection and decreasing the risk of recurrence. However, the main disadvantage of preoperative RT is its effect on wound healing.A higher complication rate has been observed when primary closure is used. Therefore, involvement of a plastic surgeon in the team may be necessary to reduce wound complications when preoperative radiation is contemplated. After preoperative radiation, 3-6 weeks interval before resection is necessary to decrease the risk of wound complications. Very long intervals between resection and postoperative radiation are not recommended.

If wide margins are obtained, additional radiation may not be needed. Often, margins are close because of the proximity of many of these tumors to major neurovascular bundles or bone. At the time of resection, surgical clips should outline the area of recurrence risk. Brachytherapy boosts should be delivered several days after surgery, through catheters placed at operation, with doses of 12-20 Gy based on margin status. Alternatively, a single intraoperative dose to the tumor bed of 10-16 Gy, based on margin status, can be delivered immediately

after resection with exposure of the area at risk, avoiding uninvolved organs. External-beam RT boosts may be an alternative to brachytherapy or intraoperative radiation: recommended doses are 10-14 Gy for close margins, 16-20 Gy for microscopically positive margins, and 20-26 Gy for grossly positive margins. Many institutions are no longer giving a boost after preoperative radiation to patients who have widely negative margins, based on local control rates that approach 95% with preoperative radiation at 50 Gy and negative margins.

Postoperative RT

Postoperative RT has been to improve local control in patients with high-grade extremity soft tissue sarcomas with positive surgical margins.When surgical resection is the initial therapy, postoperative RT choices include intraoperative radiation therapy (IORT), brachytherapy or external beam RT. RT is not a substitute for suboptimal surgical resection, and re-resection may be necessary. If the patient has not previously received RT, one could attempt to control microscopic residual disease with postoperative RT if re-resection is not feasible.

External-beam RT is delivered to large fields after surgical healing is complete (at 3-8 weeks) to doses of 50 Gy. Most institutions include the entire operative bed within that radiation field. Total doses of RT should always be determined by normal tissue tolerance. For intraabdominal or retroperitoneal tumors, this dose may be decreased to 45 Gy. An intraoperative boost may not be possible if radiation morbidity is high. If no intraoperative radiation or brachytherapy was used in the immediate operative or postoperative period, an external-beam RT boost should be added. For negative margins, an additional 10-16 Gy is recommended to a reduced field that includes the original tumor bed, based on grade and width of margins. For microscopically positive margins, an additional 16-20 Gy is recommended; for grossly positive margins, an additional 20-26 Gy is suggested.

Brachytherapy alone has been used as an adjuvant in patients with negative margins. 45-50 Gy to the tumor bed has been shown to reduce recurrence without a significant effect on wound healing.

However, brachytherapy-alone techniques require special expertise and significant experience. If brachytherapy is used as a boost, doses of 10-20 Gy based on margin are recommended; a boost dose of 10-16 Gy for close margins or 20 Gy for positive margins is recommended.

Recurrent Disease

For patients with resectable, unresectable or disseminated recurrences, the guidelines recommend the same management after biopsy, as outlined for primary disease. Preoperative RT and/or chemotherapy should be considered for recurrent disease, if not administered previously. Palliative treatment for symptom control (RT, chemotherapy or surgery) and best supportive care are potential options that oncologists should discuss with symptomatic patients. Enrollment in a clinical trial should be considered if an appropriate trial is available.

Ewing's sarcoma and rhabdomyosarcoma are typically much more sensitive to chemotherapy than are other adult soft tissue sarcomas. Adjuvant (or neoadjuvant) chemotherapy is the standard of care for adults with a diagnosis of rhabdomyosarcoma or Ewing's sarcoma. Typical regimens for small-cell pediatric sarcomas, specifically rhabdomyosarcoma and Ewing's sarcoma, include the combination of vincristine, doxorubicin, and cyclophosphamide (dactinomycin, in particular, for rhabdomyosarcoma) and the combination of ifosfamide and etoposide.

Surveillance

Patients with low-grade tumors that have been successfully resected should have a follow-up physical examination with imaging (chest/abdominal/pelvic CT) every 3-6 months for 2-3 years and then annually. Patients with high-grade tumors that have been successfully resected need more frequent surveillance. They should have a follow-up physical examination with imaging (chest/abdominal/pelvic CT) every 3-6 months for 2-3 years, then every 6 months for the next 2 years, and then annually. Chest imaging should be considered in both cases.

4. MATERIALS AND METHODS

Patients admitted for retroperitoneal sarcoma between June 2004 to November 2008 in the Department of Surgical Oncology, Government Royapettah Hospital, Chennai were taken for study.

Data were collected in all patients. Patient's age and sex were noted. Histories like abdominal mass, its duration, presence of pain and its duration, other symptoms and family history were recorded. Previous history of surgery, biopsy if any and treatment were taken. Physical examination was done to note site, size of swelling and presence or absence of metastases. Chest x-ray , CT abdomen and CT chest were taken in all patients. Histopathology, grade, and margin status were noted. Histopathology is compared with previous reports. Follow up was done once in every 3-4 months in 1-2 year and once in 6 months in 3-5 yrs and then thereafter annually once. Follow up was done with CT abdomen and pelvis and chest x ray, CT chest if required.

5. OBSERVATION AND ANALYSIS

Retroperitoneal sarcoma consisted of 0.16% of all cancers admitted from June 2004 to November 2008. Retroperitoneal sarcoma forms 10 – 20% of the soft tissue sarcomas according to Mettlin, C.Prior et al , but in this study it accounts to 9.85% of soft tissue sarcomas.

Cancers	Number	Percentage
Retroperitoneal sarcoma	21	0.16
Others	12990	99.83
Total	13011	100

Cancers	Number	Percentage
Retroperitoneal sarcoma	21	9.85
Other Soft tissue sarcoma	192	90.14
Total	213	100

In this study incidence of retroperitoneal sarcoma was seen more in males than in females in a ratio of males:females=2.5:1

Sex	Frequency	Percentage
Male	15	71.42
Female	6	28.57
Total	21	100

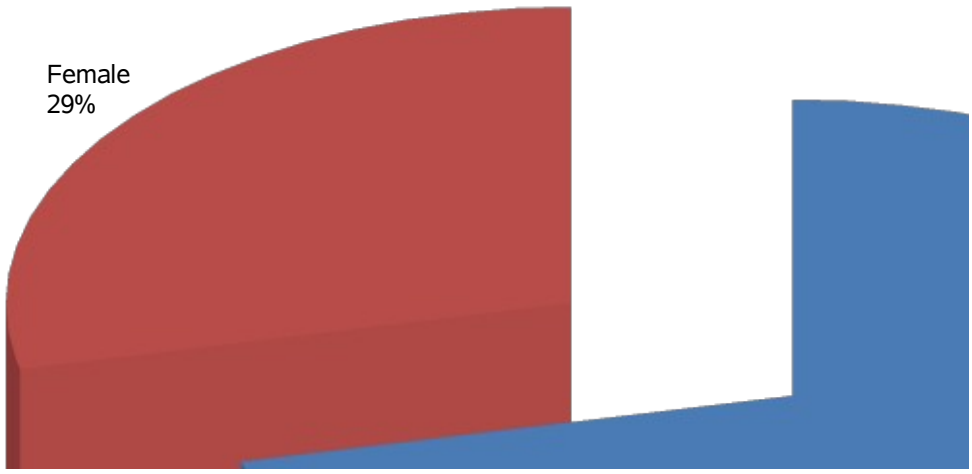
According to literature, the most common histopathologic types in the retroperitoneum are liposarcoma (40%), leiomyosarcoma (25%), MPNST, and fibrosarcoma. Approximately 55% of retroperitoneal liposarcomas are well differentiated and low grade, with tumors in roughly 40% of patients showing dedifferentiated, high-grade histologic features at primary presentation.In this study , the results are the same as that of literature with liposarcoma (52.38%) being the most common histopathology.

Histopathology	Frequency	Percentage
Liposarcoma	11	52.38
Leiomyosarcoma	4	19
PNET	3	14.3
Rhabdomyosarcoma	1	4.7
MPNST	2	9.5
Total	21	100

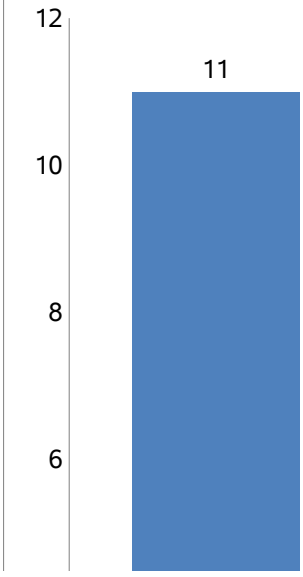
As per literature, the peak incidence is in the 5th decade of life, although they can occur in any age group. Study findings bides with literature results with peak incidence in 5^{th} decade, accounting about 33.33% of the all age groups .

Age groups(yrs)	Frequency	Percentage
<20	2	9.5
21-30	4	19
31-40	1	4.7
41-50	7	33.33
51-60	3	14.3
61-70	3	14.3
>70	1	4.7
Total	21	100

SEX INCIDENCE



Histopathology



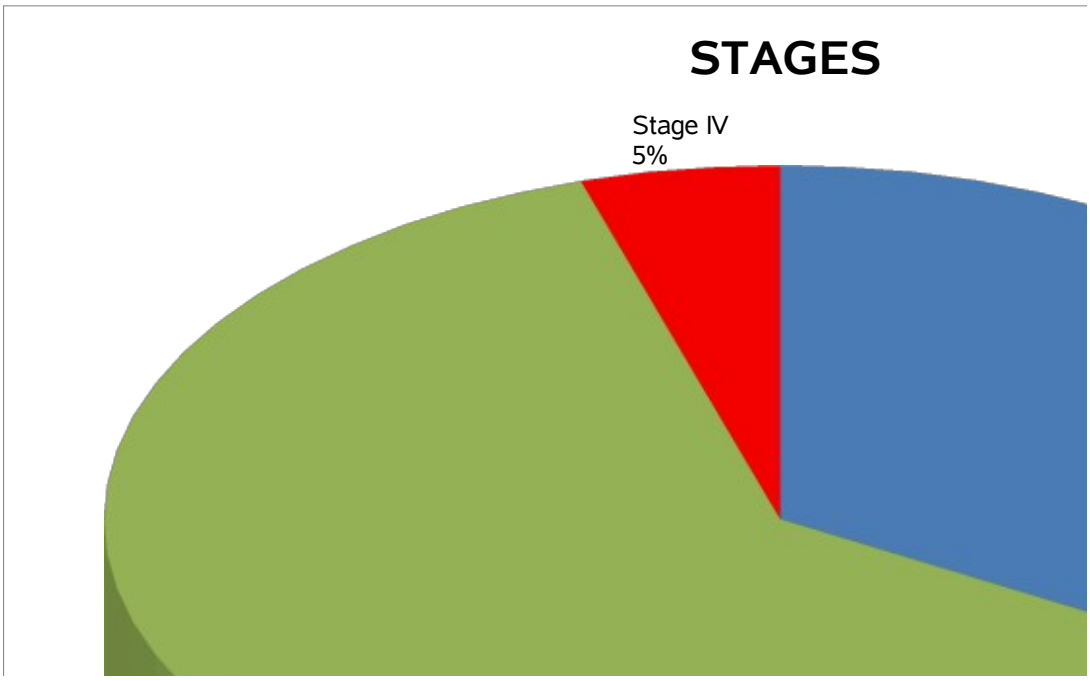
Most of the retroperitoneal sarcoma are large in size at the time of presentation and are considered as deep tumors according to the AJCC staging. All belong to T2b. In this study , the average size of the tumor at presentation is **15.19 cm**. The median size of retroperitoneal sarcoma is **25cm**.

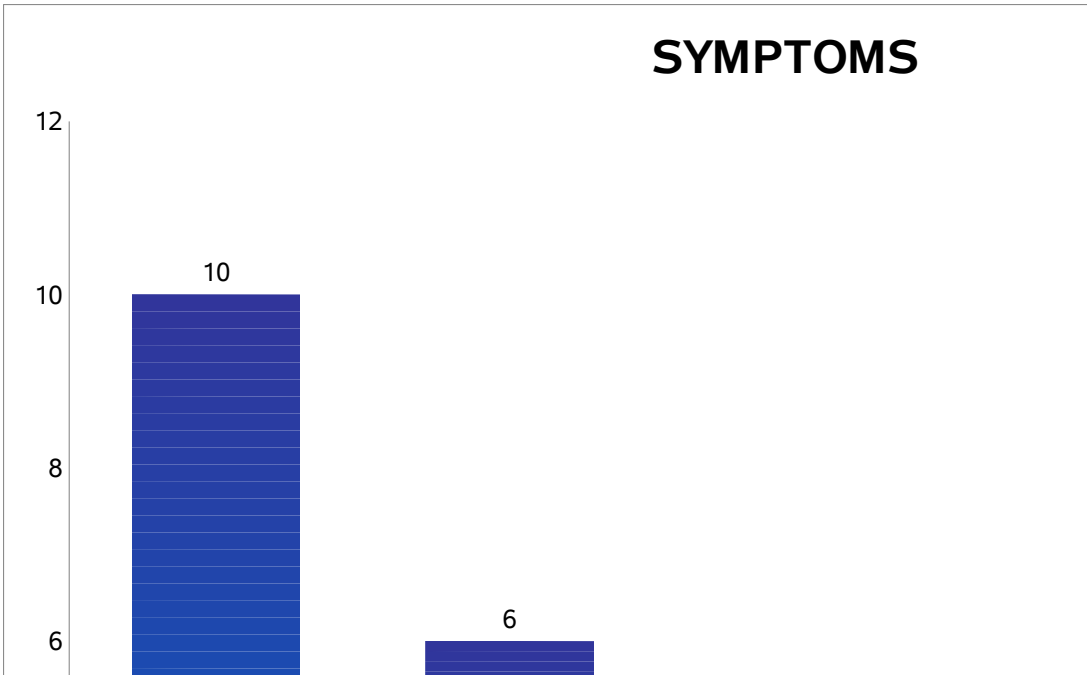
As per literature , most of the retroperitoneal sarcoma are seen in Stage III accounting to 62%.

Stage	Frequency	Percentage
IA	-	-
IB	7	33.33
IIA	-	-
IIB	-	-
III	13	62
IV	1	4.7

Only one case was found to be in Stage IV in this study.

At the time of presentation, majority were primary tumors but few of them were recurrent .


STAGES
Stage IV
5%


SYMPTOMS
12
10
8
6
10
6

Presentation	Frequency	Percentage
Primary	19	90.47
Recurrence	2	9.52
Total	21	100

Out of the two recurrent cases , one of them was a case of second time recurrence and the other 3rd time recurrence. Management and work up plan of the recurrences were managed like as for primary tumors.

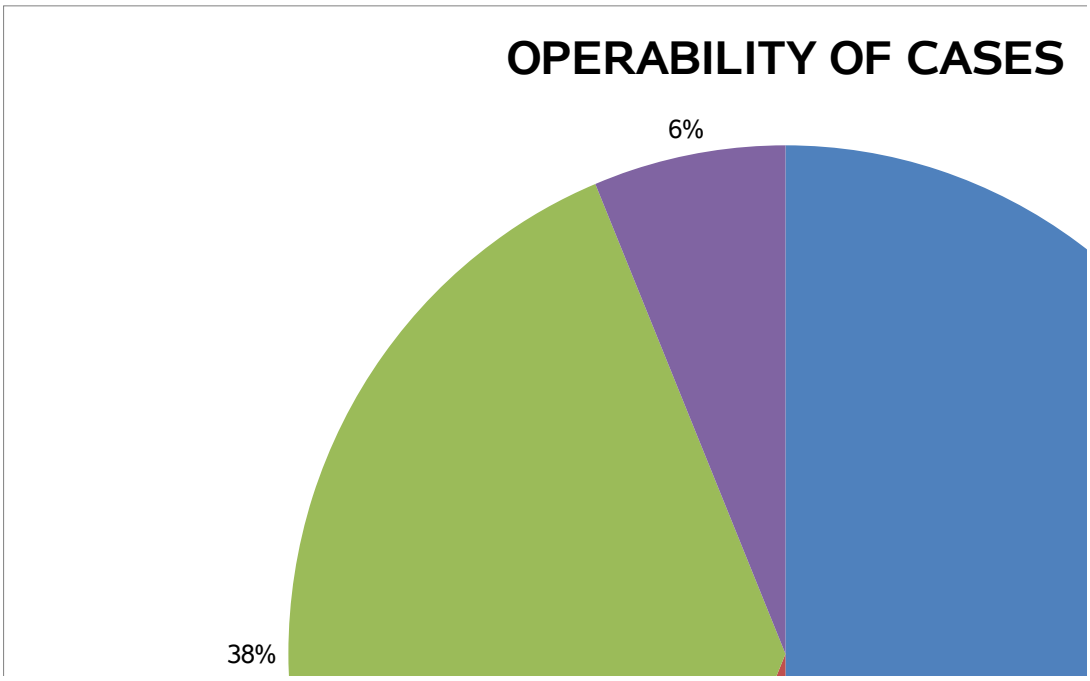
Most patients present with an asymptomatic abdominal mass. On occasion pain is present, and less common symptoms include gastrointestinal bleeding, incomplete obstruction, and neurologic symptoms related to retroperitoneal invasion or pressure on neurovascular structures. Weight loss is uncommon, and incidental diagnosis is the norm. In one report, neurologic symptoms related primarily to an expanding retroperitoneal mass were identified in 27% of patients.

Symptoms	Frequency	Percentage
Abdominal pain	10	47.6
Abdominal mass	6	28.5
GI Bleed	1	4.7
Neurologic	9	42.85

Some of the patients came with more than one complaint. Abdominal pain was the major complaint most of the patients in this study accounting about 47.6% . Neurologic symptoms was found to be high 42.85% when compared to the literature values 27%.

According to the literature it is difficult to give adequate clearance in case of retroperitoneal sarcoma because of adjacent vital structures.

	Frequency	Percentage
Complete resection	8	35
Incomplete resection	1	4.7
Inoperable	6	28.5
Metastasis	1	4.7
Chemotherapy without surgery	5	23.8


OPERABILITY OF CASES
6%
38%

Majority of the cases were resectable , but less when compared to literature results. In this study only one case was incompletely resected and in literature incomplete resection is acceptable only in case of a retroperitoneal sarcoma with well differentiated liposarcoma as the histopathology. In such cases the long term survival is significantly increased , whereas in other cases incomplete resection has the same survival rates as those without surgery.

Out of the 8 cases which had complete resection , 3 cases were positive for margins . These cases were subjected to post operative radiotherapy.

Margin	Frequency	Percentage
Positive	3	37.5
Negative	5	62.5
Total	8	100

In a study of 28 patients with liposarcomas, adjacent organ resection was carried out in more than half the cases, with partial or total resection of the kidneys in 60%, colon in 50% and adrenal glands in 35% . Although nephrectomy was performed in 60% of cases, the kidney itself was rarely involved. Nevertheless, the encompassment of the kidney and the involvement of the hilar renal vasculature make the resection of the kidney often necessary.

Adjacent organs removed	Frequency	Percentage
Kidney	6	66.66
Bowel loops	5	55.55
Total	11	100

In this study, 9 cases went in for adjacent organ resection. Nephrectomy was the most common procedure done along with resection of the tumor. Left kidney removal (55.55%) was more common than right kidney removal (11.11%). Most common bowel loops to be resected was descending colon. In most of the cases , descending colon was completely removed and distal 2/3 rd transverse colon was anastomosed to either sigmoid colon or rectum.

Majority of cases which are inoperable are due to vascular involvement(19%). Vascular structures commonly involved are inferior venacava and common iliac veins.

Blood Vessel	Frequency	Percentage
IVC	2	50
Common iliac vein	2	50
Total	4	100

In this study no vascular resection and reconstruction was done. Such cases were given post operative radiotherapy but overall survival was the same. Radiotherapy helped in local control only in few patients and tumor was progressive in some cases.

Out of the 21 cases , 12 cases (57.14%) had previous biopsy done. According to literature pre operative biopsy is only essential in case of soft tissue sarcoma of the extremity whereas not required in case of retroperitoneal sarcoma.

Pre operative biopsy is required in case of high suspicion when there is chance of the retroperitoneal tumor being a lymphoma or primitive neuroectodermal tumor where such radical resection is not at all required as they are curable by chemotherapy alone.

The grading of the post resection tumor was compared with previous biopsy results and were the same. In this study high grade tumors accounted to 66.66%.

Grade	Frequency	Percentage
High	14	66.66
Low	7	33.33
Total	21	100

According to literature most of the retroperitoneal sarcoma are low grade well differentiated tumors unlike the reports of this study.

In this study 5 cases had adjuvant and neoadjuvant chemotherapy . Especially Primitive neuroectodermal tumour(PNET) , Malignant peripheral nerve sheath tumour (MPNST) and Rhabdomyosarcoma responded well to chemotherapy. Radiotherapy and chemotherapy was also tried in inoperable and marginally positive tumors. The outcome was poor to both therapies. The chemotherapy regimen commonly used in our institute was PVCE regimen consisting of Cisplatin, Vincristine, Cyclophosphamide, Etoposide. The patients could not be followed up completely except for a few with CT chest and CT abdomen.

6. Conclusions

In this study, retroperitoneal sarcoma is a rare sarcoma accounting to 0.16% incidence of all cancers and forms 9.85% of all soft tissue sarcoma .Most of it occurs in males than in females in a ratio of 2.5:1. As with other series the age incidence is mainly in the 5^{th} decade.

The major histopathology of the retroperitoneal sarcoma is liposarcoma followed by leiomyosarcoma. Majority of the cases at the time of presentation were about 15cm in diameter and most belonged to Stage III. (62%)

Abdominal pain, discomfort and neurologic pain were the most common presenting complaints. Majority were primary tumors and only 9.52% being recurrent tumors.35% of tumors were completely resectable and 28.5% were inoperable due to involvement or proximity to vascular structures. Only one case of metastasis was reported.

62.5% of operated cases were margin negative. Most of the tumors were high grade tumors (66.66%). Most of the resections involved adjacent organ removal with bowel loops and kidney being the common adjacent organs removed. Kidney (66.66%)removal was slightly more common than bowel removal (55.55%). Vascular involvement was seen in 19% of the cases.

Only primitive neuroectodermal tumor and rhabdomyosarcoma showed good results with chemotherapy. Radiotherapy had no significant role in controlling local spread as well as on survival benefits.

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PROFORMA

Name: Occupation: DOA:

Age/Sex: IP/CD No: DOS:

Phone No: DOD:

DIAGNOSIS:

Staging:

Complaints: Abdominal pain –

Abdominal mass –

GI Bleed –

Obstruction –

Neurological symptoms –

Previous H/O malignancy:

Previous H/O biopsy:

Comorbid illness:

Treatment H/o:

H/O Recurrences:

Family H/O Cancer:

Investigations:

USG Abdomen –

Chest X-ray –

CT abdomen –

CT chest –

Biopsy - histology =

Grade =

Immunohistochemistry=

Surgery: Complete / incomplete resection:

Margin negative / not :

Adjacent organs removed :

Vascular resection :

Post-op complications:

Post-op HPE:

Radiotherapy: