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CERTIFICATE

Certified that this dissertation entitled **"MORPHOEA – A CLINICAL STUDY"** is a bonafide work done by **Dr. B.VIJAYALAKSHMI**, Post graduate student of the Department of Dermatology and Leprology and Institute of Venereology, Madras Medical College, Chennai- 3, during the academic year 2004 – 2007. This work has not previously formed the basis for the award of any degree or diploma.

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I, **DR.B.VIJAYALAKSHMI,** solemnly declare that dissertation titled, "**MORPHOEA – A CLINICAL STUDY"** is a bonafide work done by me at Madras Medical College during 2004-2007 under the guidance and supervision of **Prof. Dr. B. PARVEEN, M.D.,D.D.,** Professor and Head, Department of Dermatology, Madras Medical College, Chennai - 600 003.

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CONTENTS

INTRODUCTION

Morphoea is a benign disorder characterized by localized thickening of the skin with no known aetiological factors. Although the skin disorder resembles systemic scleroderma there are no internal organ involvement.

Morphoea is more commonly found in females and it has a characteristic morphology which differs in early and late stage. There are varying clinical types. All have characteristic distribution across the age groups. Morphoea may cause a lot of cosmetic problems which is very distressing to the patients. Sometimes it may cause growth retardations, deformity, contractures and disfigurement. There have been rare occurrence of pain, oedema, arthritis, colicky abdominal pain, visual disturbances and neurological problems in patients of morphoea. There are also interesting serological associations in morphoea.

The interesting fact about morphoea is that it resolves on its own even without treatment. So far no conclusive evidence regarding its etiopathogenisis has been proposed in spite of the advancements in the field of medicine and technology. This disease, with such peculiar manifestations has kindled the interest of dermatologist from time immemorial. Several eminent dermatologists have contributed to the knowledge of morphoea.

REVIEW OF LITRATURE

Morphoea is localized scleroderma caused by vascular sclerosis resulting in increased collagen deposition in the dermis and sometimes in deeper structures also[1].

The word morphoea is derived from Greek mythological character "Morph" who could change his body at will[2]. Erasmus Wilson is first credited for using the word to describe this lesion which he considered to be areas of vestiges of true leprosy[3]. Later Morphoea came to be used synonymously with localized scleroderma.

Morphoea has varied clinical presentations. There are also several classifications of the clinical types. The latest classification proposed by Peterson et al is as follows[4].

PLAQUE TYPE OR CIRCUMSCRIBED TYPE

MORPHOEA EN PLAQUE

KELOIDAL

BULLOUS

NODULAR

GUTTATE /LICHEN SCLEROSUS ET ATROPHICUS

ATRPHODERMAOF PIERINI AND PASINI

LINEAR TYPE

LINEAR CLASSICAL

EN COUP DE SABRE

PARRY ROMBERG SYNDROME

DEEP MORPHOEA

SUBCUTANEOUS MORPHOEA

MORPHOEA PROFUNDUS

EOSINOPHILIC FASCIITIS

PANSCLEROTIC MORPHOEA

GENERALIZED MORPHOEA

EPIDEMIOLOGY OF MORPHOEA

Incidence of morphoea is more in females.[5] The sex ratio is 3:1[5]. Whites are more susceptible.[5] Age group in which it is more common is 20 to 40 years, but it has been reported in a 1year old child and a 70 year old patient also. Morphoea is rare in children. Incidence below 10 years is 1.5% and below 20 years is 7.2%.[5] There are exceptions to this pattern. Linear morphoea has an incidence of 20% before the age of 10 years and 75% before the age 20 years .The plaque type of morphoea occurs much later in life. Only 10 % occur before 10 years and 75 % occurs between 20 and 40 years. Generalized morphoea has 80% occurrence between 11 and 50 years.[5]

The onset of morphoea is slow and insidious. The duration of morphoea is very variable, ranging from 3 to 25 years. The average is 3 to 5 years. morphoea is a self limiting disease. Therefore no conclusive studies on treatment are available.[5]

Morphoea may be associated with various other abnormalities like atrophy of fat, muscles, periostium and bones.[6] Vascular anomalies of the brain, mesentery, and kidney under lying the lesion have also been reported[7].

Most serious complications include the disturbances in bone growth under lying the lesion. Linear morphoea on limbs can give rise to limb length discrepancy and subsequent alteration in gait and height. Claw hand, hammer toes and fixed contracture of joints may also occur[8].

In a study conducted in sixty eight patients with linear morphoea 47% of them had spina bifida occulta in the radiological examination of the spine[5]. Sacralization of lumbar vertebra, presence of six lumbar segments, prolongation of transverse arches, scoliosis, rudimentary ribs, atrophic clavicle, shortened ulna, torticolis, kyphosis and contracted pelvis were some of the other bony anomalies reported.[1] Melorheostosis was seen along bones under lying linear morphoea.[43] In a rare study Prasad et al showed morphoea was associated with bone cyst.[10]

In a study conducted by Christianson on 235 patients showed that 44% of them reported with arthralgia and pain lateralized to site of lesion on the extremities[5]. In another study conducted by Singhsen et al showed 10% of

children had arthritis at a site distant to the lesion on the affected side before the occurrence of lesion.[6]

Raynaud's phenomenon was reported in 8 out of 191 patients by Christianson's study.[5] An interesting feature that has been reported is unilateral Raynaud's on side of the lesion.[11] Emotional instability, anxiety, psychoneurosis, and seizures were also reported in the same study.[5] The same study also showed 31 patients had migraine and 21 patients had colicky abdominal pain and infact 20 of these patients under went appendicectomy with no relief.

Vascular and pigmented nevi, Becker's melanosis and café au lait macules have been reported in few patients of morphoea.[5] Other autoimmune conditions like alopecia areata, vitiligo[135], icthyosis, pigmentation, nail dystrophy, hirsutism, carpal tunnel syndrome, absent pectoralis major, biliary cirrhosis and nephrotic syndrome, were some of the other features reported.[9] Rarely bullous pemphigoid and subcorneal pustulosis have also been reported in few cases of morphoea.[12]

A rare occurrence of entrapment neuropathy due to constriction by sclerosing process in subcutaneous tissue and associated hypertrichosis on lesions has been reported in a patient of linear morphoea.[13.]

Blood investigations revealed eosinophilia in both active and quiescent lesions.[14.]

Elevated immunoglobulins IgG and IgM have been demonstrated by a study conducted by Signsen et al.[6] In this, 8 out of 11 children had elevated

IgG and IgM. The same study also showed elevated ANA in most of patients with linear morphoea and also in children. He also showed that incidence of rheumatoid factor was more in children.[6.] Winkleman's study of 77 patients, showed that direct immunofluorence was positive in 35.8% of the cases.[15]

VARIOUS TYPES MORPHOEA

MORPHOEA EN PLAQUE:

This the most common type of morphoea encountered. The skin is usually smooth, shiny, thickened, and faintly purplish or mauve coloured, or rarely waxy.[16] The border is lilac coloured, well defined and circumscribed and the maximum diameter of the plaque may reach 30cm. Older lesions loose their original colour and become ivory coloured. The surface may sometimes be nodular. The hair is usually absent and there is decreased sweating.[17]There may also be loss of sensation due to the thickening of skin. The shape of the plaques may be circular, oval or irregular. There may be associated hypertrichosis.[13]They may be single or multiple or bilateral but are usually asymmetrical. The usual sites of occurrence of these lesions are the face, limbs, trunk and genitalia. They are less commonly found on axilla, perineum and nipples.[17] Sometimes vesicles, bullae, telengectasia and haemorrhages may also be seen on the lesion.[18]

The onset is usually insidious and sometimes rapid with associated oedema and erythema.[17] Sometimes pigmentation may precede the appearance of lesion.[5] Rarely pain may occur before the onset of lesions. Migraine or

colicky abdominal pain may be associated with lesions occurring on the face and abdomen respectively.[5]

KELOIDAL MORPHOEA:

This is a rare variant of morphoea. This resembles a keloid and it shows greater depth of inflammation, but limited to dermis.[19] There is increased fibrosis and collagen deposits. This type of morphoea is mostly seen associated with systemic scleroderma.

NODULAR MORPHOEA

These can occur along with plaque type of morphoea. The surface is extremely nodular.[20.]

BULLOUS TYPE

Blisters may be seen in morphoea. This may be due to lymphatic dilatations or vascular sclerosis causing obstructions. Eosinophils capable of releasing major basic proteins causes subepidermal lysis and produces bullous lesons.[22]. Similar condition may also be seen in Lichen sclerosis et atrophicus.

GUTTATE MORPHOEA (LICHEN SCLEROSUS ET ATROPHICUS/WHITE SPOT DISEASE)

Guttate means rain drops. Since these lesions are also small like rain drops, chalky white and size varying from 1 to 10 mm these are called guttate morphoea. It is a very superficial type of morphoea .These lesions also have a well defined lilac coloured border and are seen commonly on trunk, chest,

neck, shoulder and extremities. Multiple lesions can be seen on the same person.[28]

As per the classification of Peterson[4], Lichen sclerosus et atrophicus is also considered to be type of guttate morphoea. Bizzero in his study also considers it to be a type of guttate morphoea.[23] Certain authors like K.G. Bergstorm consider it to be different because of early development of epidermal atrophy and follicular dellings and variations in the histopathology.

LICHEN SCLEROSUS ET ATROPHICUS:

The lesions are porcelain white and have follicular dellings (widened pilosebacious orifices) on them. They have specific HLApredispsition.[24, 25]

Female related HLA includes :

MHC I - HLA -B40, HLA -A29, HLA -B44.

MHC II - DQ7, DQ8, DQ9.

Males related HLA includes :

- DR11, DR 12, DQ7

Skin is atrophied and numerous lesions are seen on trunk, limbs and genitalia.[23.] Number of autoimmune disease are associated with Lichen sclerosis et atrophicus[26]. Borrelial infection have been implicated in development of these lesions.[90]

These lesions may also follow the Blasckho's line. Lichenification may be seen surrounding the lesion due to frequent rubbing. Occasional

telengectasia, purpura, or bulla may be seen on the lesion. In later stages, they may become wrinkled and depressed. Scarring alopecia may occur on the scalp.[28] Involvement of superior oblique muscle of the eye may give rise to diplopia.[29.]

Anogenital lesion in women and men may occur at 45 to 60 years of age. Rarely female children may also have anogenital lesions. Hormonal influence may cause of such lesions.[86] Constant friction on these surfaces may give rise to raw areas which may later heal to produce flat and glistening lesions. Patients usually complain of soreness and dyspareunia. A late complication may be resorption of labia, vulva, and clitoris leading to narrowing of vaginal introitus. Normal pregnancy and delivery have occurred despite this problem.[30]The male counterpart of this disease is called balanitis xerotica obliterans[23] which presents with recurrent balanitis phimosis and painful erection.

ATRPHODERMA OF PIERINI AND PASINI

This is a primary abortive form of morphoea in which indurations fails to occur[31]. It is an uncommon superficial form of localised morphoea in which there is an oval, hyper pigmented or slate grey atrophic plaque on the trunk. The plaques may be multiple and the centre is usually depressed and the border is described as typically being cliff-drop like.[32]. Some authors consider it to be an optical effect because of the difference in colour.[33] It occurs much earlier in life around 10 to 20 years of age and has a protracted course. It also lacks the typical violacious border. It may coexist with other types of morphoea, suggesting that this could have resulted from transformation of these lesions.[35]

Another peculiar feature of this morphoea is that, atrophy occurs much before sclerosis.[36] Borrelia burgdorferi has also been implicated in the development of this lesion.[90]

LINEAR MORPHOEA (SYN. CLASSICAL LINEAR MORPHOEA)

Localised scleroderma occurs in linear bands in this condition. They are usually unilateral or rarely bilateral. The extremities are usually affected and lower limbs are much more commonly affected. These linear lesions may also occur on anterior aspect of thorax, thighs and buttocks. Homolateral lesions involving one arm and same side leg have also been reported. Rarely one half of the body may be affected.[37]

These lesions usually lack the usual violacious ring but they may be seen in the advancing boarders. They may also follow the lines of Blaschko[38]. These distributions along lines of Blaschko shows that the tendency to develop scleroderma is predetermined during embryogenesis with formation of clones of vulnerable cells which on appropriate trigger may transform to develop full blown lesion.[39]

Patients usually complain of pain, arthralgia or oedema before the onset of the lesions.[5]The underlying bones and muscles may be involved in 20% of cases leading on to severe growth disturbance and limb length discrepancy. When these lesions cross a joint it can give rise to contractures.[40] On radiological examination, the underlying involved bone may have a characteristic picture of wax flowing out of the candle due to cortical hyperostosis.[41] This peculiar condition is called melorheostosis. This is

considered to be developmental anomaly. A sclerotome[147] supplied by a spinal nerve may be affected in this condition This is a painful condition seen mainly in children and presenting with asymmetrical contractures and thickening of the overlying skin.[42] Any surgical correction may further worsen the condition.

Rarely linear scleroderma may be associated with overlying hypertrichosis.[44.] Ulcerated dystrophic calcinosis[45] and nodular morphoea[46] may also be seen on linear morphoea. Sometimes bands[47] of lesions occur circumferentially around limbs, fingers or breasts, resembling an ainhum.[48,50,] The tissues distal to these bands may be oedematous and depigmented.[49] Fibrosis may spare areas with in a linear morphoea producing skip lesion. Linear morphoea of lower limbs is more commonly associated with spina bifida occulta.[5]

FRONTOPARIETAL MORPHOEA: (SYN. EN COUP DE SABRE)

En coup de sabre in French means a strike with a sword. Atrophy of skin is usually present and the skin over the affected area is usually contracted and firm in the early stages. Later an ivory coloured sclerotic plaque develops and there may be rarely overlying telengectasia and hyperpigmentation of border. Later a depressed groove appears on the frontoparietal region, extending on to the scalp. In the scalp there is alopecia which in some cases may be preceded by bleaching of hair.[54]The groove may extend further down on to the cheeks, nose, upper lip and gums. In severe case it may involve the chin and neck also. It may affect the gingiva and jaws also leading to alteration in arrangement, spacing and direction of teeth.[52] Tongue may be atrophied or grooved. There may be atrophy of one half of the face leading on to facial asymmetry.[54]

Rarely frontoparietal lesions may be bilateral, trilinear or may follow Blaschko's lines.[53] There may be associated morphoea lesions on other sites of the body or there may be total atrophy of one side of the body either homolateral or contralateral or rarely both sides.[56,57] The underlying bone can also get affected. There may be EEG abnormalities showing dysarrythmias under the affected site.[28] Variety of ocular lesions like enophthalmous, lid ptosis, extroccular muscle weakness, iris atrophy , heterochromia iris , atrophy of fundus ,loss of eye lashes (along the line of involvement) and eye lid oedema may be the presenting feature.[58,59] Rarely ossification may occur in later stages.[57]

PARRY ROMBERG SYNDROME

Hemiatrophy of Parry Romberg is similar to frontoparietal morphoea but there is usually no cutaneous sclerosis. It usually begins as hyperpigmentation on the face. The skin is less bound down to the deeper structures and it is usually seen along the distribution of the trigeminal nerve.[56.]

DEEP MORPHOEA

MORPHOEA PROFUNDUS

Whittaker et al described this morphoea, in which all layers of the skin are involved.[61]They appear as solitary fibrotic plaque on shoulders, back, neck, or paraspinal areas. The skin is invariably hyperpigmented .Osteoma cutis develops very often in these lesions.[62] The skin is bound down and borders are usually ill defined. They have a cobble stone or pseudocellulite appearance. The "groove sign" (a depression along the course of a vein between muscle

groups) may be evident.[28.] In subcutaneous morphoea, the subcutaneous plane alone is involved and there no signs of inflammation on the skin. Flexions contractures are more common in this type.[21]

DISABLING PANSCLEROTIC MORPHOEA OF CHILDREN

This a rare forms of morphoea. Dermis, fat, fascia, muscles and even bones are sclerosed before the age of 14 years. It may develop from linear morphoea or de novo. Trunk, extremities, scalp, and face may be involved .The finger, soles and toes are exceptionally spared.[63] The patient may walk on tip toes due to contractures of Achilles tendon. The onset is usually acute and there may be associated arthralgia and stiffness. The intense pain may be due to involvement of cutaneous nerves.[64] Oesophageal, pulmonary and periodontal changes may be seen. Flexion contractures, osteoporosis and bone changes are some of the common complications seen.

Electromyogram is usually abnormal. Histopathological changes in muscle may be seen .Creatinine phosphokinase level is normal. Serological investigation shows elevate ESR, eosinophilia and hypergammaglobulinemia. Treatment with PUVA and cyclosporine have been useful. With out treatment this condition is progressive and occasionally fatal.

GENERALIZED MORPHOEA (SYN. GENERALIZED SCLERODERMA)

In this condition there is generalized sclerosis of the skin. There is no systemic disturbance. It occurs between 30 to 40 years of age.[65]A rare incidence has been reported below 11yrs and above 50 years.[5] Females are

more commonly affected more than men. The sex ratio is 3:1.[5] On set is usually insidious. Individual plaques look like localized morphoea. It usually begins in the trunk. The hands may resemble the tumid phase of systemic scleroderma. Spindling of fingers may be seen. Flexion contractures have also been reported[5]. Rarely the whole body from head to foot may be involved.[66] There may be dyspnoea when chest wall is involved. Respiratory failure has been reported due to inter costal muscles involvement. The face may be shiny and brown, and expressionless due to indurations of the under lying skin and there may be restricted mouth opening. Raynaud's phenomenon have also been reported in few cases.[5] Whitlows following trauma have been reported.[67]Non pitting oedema may precede the development of morphoea. Bullous, keloidal or nodular type may coexist with generalized morphoea. Acral myofibromas may occur in few cases.[67] Overall hyperpigmentation may be the presenting symptom.[5]The skin lesions of generalised morphoea may be superimposed by keratosis or calcinosis.[65]

Contracture of joints, thinning of limb and soreness may be the presenting complaints in acute phase of the disease. Joint pains were reported in 50% of cases[5]. Rheumatoid arthritis, polymyositis and sick sinus syndrome may coexist along with this condition.[68] Sometimes intractable pain, infection and atrophy of the limbs may occur in this condition. Squamous cell carcinomas have occurred in long standing case of morphoea.[1]

Generalised morphoea may also coexist with lichen sclerosus et atrophicus.[133] Subcutaneous involvement of lesions occurring in generalised morphoea is more inflammatory than superficial morphoea. Rarely

subcutaneous involvement in generalised morphoea may occur with systemic eosinophilia.[114]

EOSINOPHILIC FASCIITIS: (SYN. SCHULMAN'S SYNDROME)

Although it is a distinct entity some authors consider it to be a variant of morphoea.[69.] It forms one end of spectrum of linear morphoea and the other end of systemic scleroderma. It may be associated with other connective tissue disorders. It occurs both in children and adults. Males are more commonly affected. There is acute pain before the onset of the lesions, which is followed by swelling and tenderness of the distal parts of the limbs and later there is induration.[70]There is limitations of movement of feet and hands. Occasionally the lesions can occur on face or abdomen and there may be superficial blistering or haemorrhage on them. Trauma or strenuous exercise is considered to precipitate this condition. There is no Raynaud's phenomenon, internal organ involvement or history of previous infections in this condition.

Eosinophilic fasciitis like syndrome have also been reported after ingestion of L-Tryptophan.[71] In addition to the usual feature of muscle weakness, muscle enzymes are also elevated. Enhanced type I procollagen gene expression in the skin has been demonstrated.[72] In another similar condition called eosinophilia-myalgia syndrome, there may be dyspnoea, oedema, arthralgia and rashes in addition to the above features.[73.] This condition is also precipitated by ingestion of L-Tryptophan.[74]

Histopathological examination of eosinophilic fasciitis may be similar to morphoea. There may be raised ANA titre, hypergammaglobulinemia and eosinophilia.[75]This condition also resolves on its own.[72.]

ASSOCIATIONS OF MORPHOEA

Although morphoea is a localised condition with out any systemic involvement, there are many interesting associations with morphoea. Arthralgia,[5]unilateral Raynaud's[11] migraine and colicky abdominal pain[5] have been reported. Spina bifida was found in some of the cases of linear morphoea.[5,96,101] Sacralization of lumbar vertebra, presence of six lumbar vertebrae, prolongation of transverse arches, kypohosis, scoliosis, lumbar intervertebral disc prolapse, rudimentary clavicle, torticolis, atrophic clavicle, absent pectoralis muscle, contracted pelvis, shortened ulna and deformities of feet and toes were other bony defects described.[5] Warty vascular pigmented naevi, usually on the same side of morphoea, lichen planus[146] café au lait spots, alopecia areata, vitiligo,[135] generalised icthyosis, pigmentation, dystrophy of nails and hirsutism have also been reported in few patients.[1] Children may be intellectually precocious.[5] Variants of morphoea occurring later in life may also be associated with tissue calcification. Hair loss and beaking of nose were also seen in few studies.[9] Bullous pemphigoid, subcorneal pustulosis and primary biliary cirrhosis have also been reported as associations.[12] Localised morphoea and Lichen sclerosus et atrophicus have occurred in same patients.[136]

No systemic changes were found in morphoea except for oesophageal changes in 27% of cases.[5] systemic lupus erythematosis[107], progressive scleroderma, dermatomyosites[110], carpal tunnel syndrome[15], myasthenia gravis,

nephritis, elastosis perforans serpiginosa, discoid lupus erythematosis, rheumatoid arthritis[146] and mixed connective tissue disorder were other autoimmune disorders seen in morphoea.[108] Silicon implants into breast for augmentation may also cause morphoea.[104] Morphoea like features have also followed carcinoid syndrome.[105] There is no relation between internal malignancy and morphoea. Squamous cell carcinoma[1] have occurred in long standing cases morphoea.

ETIOPATHOGENISIS

Pathogenesis of morphoea is still a mystery. Various studies have been conducted and numerous theories have been put forward to explain the evolution of morphoea. The basic fact is that there are increased fibroblasts, and increased production of collagen.

There is always a triggering factor for production of sclerosis. A vascular injury may be the primary insult promoting migration of mononuclear cells across the damaged endothelial cells. The endothelial cells promote increased production of platelet derived growth factors and their receptors.

There is increased production of Transforming growth factors-β (TGF-β), which may increase connective tissues growth factor[76] (CTGF) gene expressions and promote fibrosis. There are also increased production of interleukin 2 (IL 2)[77] receptors, IL6 receptors, CD4, CD8, CD28, CD30, TNF-α[78], soluble vascular cell adhesion molecules (VCAM-1), E selectin[79] and endothelial cell antibodies. All these promote fibrosis. Such changes have also

been reported in graft versus host disease and idiopathic thrombocytopenic purpura.[80]

A possible pathogenic role of fibrillin –1 in localised sceleroderma has been demonstrated by isolation fibrillin –1 antibody[82,83] in the serum of patients. The progenitor cell antigen CD34[84] identified in a subpopulation of dendritic cells are decreased in localised scleroderma, proving that a loss of immunomodulations may result in pathogenesis of morphoea. In melorheostosis of bone of, TGF-β is increased causing proliferation of periosteal fibroblasts.[77]

Connective tissue growth factor (CTGF) has the ability to induce TGF-B and ultra microscopy and RNA analysis shows that CTGF m RNA is increased in lesional cells. TGF-β & CTGF form a loop that induce and sustain fibroblast proliferation and extracellular matrix production.[83] Antibody to U1RNP has also been implicated in development of morphoea.[81]

ETIOLOGICAL THEORYS

Various theories and precipitating factors have been proposed for the development of morphoea. Few of them are as follows.

GENETICS

There is increased incidence of morphoea in family members of patients.[85] Monozygotic twins have had morphoea. So far HLA predisposition has not been elucidated. There are increased organ specific antibodies in relatives of patients.[86]

TRAUMA

Trauma preceding the development of lesions have been reported in several studies.[87] Trauma may trigger vascular sclerosis and lead on to cutaneous sclerosis.[88] Surgical trauma has caused morphoea following surgery for atriovenous fistula and rhinophyma.[89]

VACCINATION

Vaccination for B.C.G,[93] varicella zoster[94] and tetanus[90] have triggered morphoea.

Injection

Injection vitamin K[91] has produced lesions around the hip girdle. The characteristic distribution is called gun-belt and holster sign.[91] In several other studies shows injections whose nature was unknown have also precipitated morphoea probably due to trauma.

HORMONAL CAUSES

Pregnancy has precipitated morphoea in many females.[86] Extract of pratharmone injections in rats have produced sclerodermatous changes and calcium deposits in skin and decalcification of bones.[93] Lichen sclerosis et atrophicans in prepubertal girls have resolved after puberty. Improvement of lesions in menopausal women after hormonal therapy further supports this view.[86]

METABOLIC CAUSES

Phenylketoneuria has been implicated in development of morphoea.[92] Low phenylalanine diet has resulted in resolutions of these lesions. Diabetes mellitus has also produced scleroderma like changes.

INFECTION

Several infections have been implicated in development of morphoea. Important among them is Borrelia burgdorferi.[94] Polymerised chain reactions have detected DNA of these bacteria in lesional skin. Generalized morphoea has followed measels.[95]

DRUGS

Several drugs have been implicated in development of morphoea. They include penicillamine[98], bromocriptine[99], hydroxytryptophan[109], carbidopa[104], pentazocine,[96] docetaxal[100] and bleomycin.[97] Isolated limb perfusion with melphalan[104] have caused morphoea in few patients on limbs[103]. Among this penicillamine have been used as a therapy for morphoea and has induced morphoea in few patients. There has been spontaneous improvement after stopping these drugs. Valproic acid used in treatment of seizure has also triggered morphoea.[104]

RADIOTHERAPY

Morphoea at the sites of radiotherapy have been reported. Breast cancer treated with chest wall irradiation has later on lead to development morphoea in the same region and also in the calf region.[105a]

MISCELLANEOUS CAUSES

Exposure to epoxy resins, vinyl chloride, organic solvents and pesticides have produced scleroderma like changes.

NEURAL THEORY

Linear morphoea of lower limbs have been associated spina bifida occulta.[5] In a study conducted by Rubin et al 17.3% to 33% patients had spinabifida occulta in lumbosacral spine.[101] This defect could have damaged the spinal cord, by trauma or adhesion of cauda equina to the defect may give rise to trophic changes of lower limbs resembling the changes seen on morphoea. This view supports the theory that nerve damage is the important cause of development of morphoea.[102]

Segmental morphoea or linear morphoea that usually follows the course of peripheral nerves further supports this theory that injury of these nerves could have triggered morphoea. The prolongation of sensory chronaxie in systemic scleroderma further strengths this theory.[105]

IMMUNOLOGICAL THEORY

Other autoimmune conditions have also been associated with morphoea.[86]This association suggests that morphoea could also be an autoimmune disorder. Pernicious anaemia, hypothyroidism, thyrotoxicosis, alopecia aereata, diabetes and vitiligo[135] have been reported to occur in association with morphoea.[108] Antithyroid antibodies[108], ANA[6] and antiparietal

antibodies[108] have also been isolated in considerable cases of morphoea. Morphoea like lesion have been reported in graft versus host reactions.[80]

Deposition of immunoreactive substances in lesional skin further strengthens this theory. Winkle man et al has demonstrated positive direct immunofluorence in 24 out of 77 patients(35.8%) in his study.[15]

There are many studies that show morphoea has followed or coexisted with other autoimmune diseases like systemic lupus erythematosus[111,112] and progressive systemic sclerosus.[110]

Presence of hypergammaglobulinemia[6], positive rheumatoid factor[6] and L.E[110] cells are additional supports for immunological theory.

INVESTIGATIONS

Anaemia is seen in several patients of morphoea.[28.] ESR is raised in many patients with active morphoea.[137] Eosinophilia (>400cells/cumm) is also present in many of the patients.[14] Antihistone antibodies [115] were positive in 25% of plaque type of morphoea and 32% of linear morphoea. Anti single stranded antibodies to DNA[116] are frequently found, in generalized morphoea (75%), linear morphoea (53%) and plaque type (27.3%). Rheumatoid factor[117] is especially raised in children with linear morphoea.[6] Hereditary deficiency of C2 has been demonstrated by Hulsman in study.[118] Organ specific antibodies were also found in relatives of morphoea.[86] Idiopathic thrombocytopenia were present in many patients, which improved with corticosteroid therapy were recorded[119]. Antinuclear[28] antibodies were also positive in 40% of patients and

showed both homogenous and speckled pattern. Procollagen peptide I and III[121], antibody to topoisomerase II[28] and antibody to Zn/Cu super oxide dismutase[28]have been recently reported to be raised in patients of morphoea.

Severe hypocomplimentemia, eosinophilia, raised ESR, and anti DNA are useful in judgement of activity of morphoea.[120] The progression to systemic scleroderma is assessed by presence of Anti ku antibodies.[123]

ULTRASONOGRAM

Ultrasound scanning of the lesion with 20MH B mode pulse measures the thickness of the morphoea[122]and is also useful in assessing the prognosis.

RADIOLOGICAL EXAMINATION

Radiological examination of skull and limbs shows underlying growth arrests. A peculiar condition called melorheostosis[41] of bones showing cortical hyperostosis can be made out. Shortening of limbs, indurations of skull and other bony abnormalities like sacralization of lumbar vertebra, contracted pelvis, rib anomaly, atrophy of clavicle, spina bifida occulta, shortened ulna, kypohosis, and scoliosis and bone cysts can be made out.[1,5]

MRI OF BRAIN

Frontoparietal morphoea may show cortical atrophy, ventricular dilatation, calcification of leptomenninges or anomalies of intracranial vasculature.[28]

MRI IN EOSINOPHILIC FASCIITIS

This shows oedema of subcutaneous tissue and increased signal intensity on T2- waited images and contrast enhancement of the facial planes[28].

ELECTROENCEPHALOGRAM

Dysarrythmias in EEG underlying the lesional skin have been reported[24]in frontoparietal morphoea.

DIRECT IMMUNOFLUORENCE

This shows IgM, IgG, andC3 deposits were seen around the blood vessels of the sclerosed skin.[15]

HISTOPATHOLOGY

It is difficult to differentiate the different clinical types of morphoea by histopathology but the difference in depth of involvement can be appreciated. Severity can be graded according to the depth of involvement.

Based on the evolution of morphoea they can be graded as early, intermediate and late.

EARLY STAGE

Biopsy of the specimen in early inflammatory stage especially from violacious border shows dense lymphocytic inflammatory infiltrate in dermis and around blood vessels. Collagen bundle are only slightly thickened. In later

stages vascular changes are seen in subcutaneous tissue also. Endothelial swelling are seen in the vessal walls.[125]

INTERMEDIATE STAGE

A much more pronounced inflammatory infiltrate is seen in dermis and subcutaneous tissue extending upwards to surround the eccrine glands. Trabeculae subdividing the subcutaneous fat are also thickened due to deposition of newly formed collagen. Collagen is usually composed of fine, wavy fibres which are arranged in bundles and stain faintly with eosin-haematoxylin stain.[124]

LATE STAGE

Biopsy from the centre of the plaque or from an old plaque shows no inflammatory infiltrate in dermis except for few in subcutaneous tissue .The epidermis may be atrophied and there is loss of rete ridges. The collagen in reticular dermis appears thickened, closely packed, hypocellular and stain more deeply with eosin than normal skin. In papillary dermis the collagen is more homogenous.

The eccrine glands are markedly atrophic and are conspicuous by absence of adipocytes surrounding them and are surrounded by newly formed collagen. There are very few blood vessels seen with in the sclerotic area. The walls of vessels are also fibrotic and the lumen is narrowed. Special elastic stains show thick elastic fibres lying parallel to the collagen strands. Similar process is also seen in subcutaneous tissue.[124]

DEEP MORPHOEA

The fascia shows increased fibrotic process as seen in subcutaneous tissue and muscle fibres appear vacuolated and separated from one another by oedema and there is focal collection of inflammatory cells.[125]

BULLOUS MORPHOEA

This is a rare type of morphoea. There is a sub epidermal separation due to oedema because of lymphatic obstruction.

ATROPHODERMA OF PASINI AND PEIRINI

Histopathological changes are minimal, mild and non-specific. There is only minimal thickening of collagen bundles and scattered inflammatory infiltrate. Older lesions show no inflammatory infiltrate but only thickened collagen tightly packed in deeper layers of dermis. Indurated area show homogenous, hyalinized, hypertrophic collagen. The lesions are usually present in the back. The normal collagen in the back is usually thick. Therefore for comparison sake either bit of normal skin from same side or from the opposite side has to be included in biopsy.[126]

EIOSINOPHILIC FASCIITIS

A deep wedge shaped biopsy from a lesion shows thickened fascia which appears homogenous and permeated by mononuclear cells. In some cases infiltrate is predominantly eosinophils.[28]The under lying myofibrils in the skeletal muscles shows degeneration and severe inflammation with a increased component of eosinophils and focal scarring.

The adipose tissue shows no significant changes except for fibrous septa separating fat lobules which are thicker, paler, homogenous and hyalinized. Sometimes the collagen in lower reticular dermis and subcutaneous tissue also appears pale and later they may get horizontally oriented merging with the fascia.[127]

LICHEN SCLEROSUS ET ATROPHICUS

Epidermis is atrophic with follicular hyperkeratosis. The basal layer of cells shows hydropic degeneration.[129]The stratum malpighii also shows atrophy. Rete ridges are completely absent in most of the lesions. Appearance of keratotic plugging is associated with disappearance of appendageal structures. Keratotic plugging is also absent in mucosal lesions.[130] There is also mild inflammatory infiltrates in the middle of the dermis. There is pronounced oedema and homogenization of the collagen in upper dermis.[131]The collagenous fibres also appear swollen, glassy and contain only few nuclei. They stain poorly with eosin stain and elastic fibres are very sparsely present.[132]

ELECTRON MICROSCOPY

Endothelium of blood vessel shows vacuolizasation. There is also reduplication of the basement membrane. Mononuclear cells, pericytes and fibroblasts show enlarged endoplasmic reticulum which confirms their increased activity. Perivascular inflammatory infiltrate precedes the stage of fibrosis. There is increased synthesis of collagen with smaller diameter of 50

nm or less. The normal diameter is 70 to 140nm. The ratio of Type I & Type III collagen is unaltered.[133]

HISTOCHEMICAL STUDY

There is excessive production of ground substance and fine collagen fibres. There is increased amount of PAS positive diastase resistant material in areas of homogenization of collagen. The glycosaminoglycons show alteration in structure. Chemical analysis shows increased hexosamines and hexoses bound to collagen fibres. Sugars attached to the collagen give the homogenous appearance in eosin- haematoxylin stain.[134.]

TREATMENT

There is no specific treatment for morphoea since it resolves on its own. Various topical, intralesional and systemic therapies have been tried. Some of the topical application includes steroids,[138]calcipotriol[138] and topical photodynamic therapy.[139]

Intralesional steroids[138] have also been tried. Some of the systemic therapies tried includes PUVA,[140] methotrexate,[141] grisiofulvin,[142] phenytoin,[143] doxycyclin,[138] etretinate,[138] chloroquine,[138] pencillamine,[98] steroids[144] vitamin E[146] and cyclosporine.[145]

AIMS OF THE STUDY

1 To study the incidence of Morphoea in Government General Hospital, Chennai during the period of two years between September 2004 and September 2006

2. To study the incidence of various types of Morphoea.

3. To study the sex wise distribution.

4. To study the age wise distribution.

5. To study the commonest site of lesions.

6. To study the main presenting complaints.

7. To study the precipitating factors.

8. To study the relevant serological abnormalities.

9. To study the associated autoimmune disorders.

10. To study the other associated anomalies.

11. To correlate the Histopathological findings with various types of morphoea.

12. To study the incidence of morphoea in relatives.

MATERIALS AND METHODS

All the patients attending the outpatient department of Government General Hospital Chennai during the period between September 2004 and September 2006 were screened and patients with morphology suggestive of morphoea were enrolled in the study. There were forty one patients who had morphology similar to that of morphoea. A detailed history as given in the proforma was elicited. Various presenting complaints like loss of sensation, hair and sweating over the patch and other problems like contractures, shortening, head ache, joint pains and seizures were also recorded. Biopsy was done on all patients. All patients whose biopsy was similar to that of morphoea was subjected to various other investigations. Total count, differential count, haemoglobin, erythrocyte sedimentation rate, absolute eosiniphil count, blood sugar, blood urea, urine albumin, urine sugar, liver function test, Antinuclear antibody titer, Rheumatoid factor and 'C' reactive proteins were tested. Radiological pictures of spine of patients with linear morphoea of the lower limbs were taken. Relevant radiological examination of the limbs and skull were also taken. Opinion of Neurologist and Ophthalmologist were obtained in patients of frontoparietal morphoea. All the data were compiled and inference drawn.

OBSERVATIONS AND RESULTS

INCIDENCE

Of total 30,000 patients attending Skin O.P Government General Hospital, Chennai, during the period between September 2004 and September 2006, total number of patients with morphoea was 41.

Incidence of morphoea was 1per 1000 or 0.14%

TABLE - 1

Total no. of patients attending Skin O.P in Govt. Gen. Hosp. (Sep 2004 to Sep 2006)	30,OOO
Total no. of patients with morphoea	41
Incidence of morphoea	0.14 %

DIFFFERENT TYPES OF MORPHOEA AND THEIR INCIDENCE

Percentage of different types are as follows:

Linear – 36%

Plaque – 34%

Frontoparietal –20%

Generalized –5%

Mixed –5%

TABLE – 2

S. No	Clinical types	No of cases
1	Linear type	15
2	Plaque type	14
3	Frontoparietal	9
4	Gen. morphoea	2
5	Mixed (plaque and linear)	2
	Total	41

AGE WISE DISTRIBUTION

Youngest person was 4 year old female child

Oldest person was 49 year old woman

Incidence of morphoea peaked between 10 to 25 years

Incidence below 10 years was 9%

Incidence below 20 years was 48%

TABLE – 3

Age in years	Females	Males	Total
0 - 4	0	1	1
5 - 9	3	0	3
10 - 14	6	2	8
15 - 19	7	1	8
20 - 24	3	5	8
25 - 29	0	1	1
30 - 34	2	0	2
35 - 39	3	0	3
40 - 44	0	1	1
45 - 50	4	2	6

AGE WISE DISTRIBUTION OF PLAQUE TYPE

Incidence below 10 years was nil

Incidence between 20 to 50 years was 34%

TABLE - 4

Age	Male	Female	Total
0 - 9	0	0	0
10 - 19	1	4	6
20 - 29	1	1	2
30 - 39	0	3	2
40 - 49	3	1	4
Total	5	9	14

AGE WISE DISTRIBUTION OF LINEAR TYPE

Incidence below 10 years was 2%

Incidence below 20 years was 14%

TABLE - 5

Age	Male	Female	Total
0 - 9	1	0	1
10 - 19	0	6	6
20 - 29	2	-	2
30 - 39	0	3	3
40 - 49	1	2	3
Total	4	11	15

AGE WISE DISTRIBUTION OF GENERALIZED TYPE

Incidence between 11 and 50 years was 2%

TABLE - 6

Age	Male	Female	Total
0 - 9	0	0	0
10 - 19	0	1	1
20 - 29	0	0	0
30 - 39	0	0	0
0	1	1	1
Total	0	2	2

AGE AND SEX WISE DISTRIBUTION FRONTOPARIETAL

Incidence in males and females are equal

Incidence between 10 to 30 years was 20%

TABLE - 7

Age	Male	Female	Total
0 - 9	0	0	0
10 - 19	2	2	4
20 - 29	2	2	4
30 - 39	0	0	0
40 - 49	0	0	0
Total	4	4	8

AGE AND SEX WISE DISTRIBUTION OF MIXED TYPE

Incidence of mixed type of morphoea was 5%

TABLE - 8

Age	Female	Male	Total
0 - 10	2	0	2

DURATION OF EVOLUTION OF MORPHOEA

Average durations of evolution of morphoea was 2 to 3 years

TABLE – 9

Evolution period in years	No. of patients
0 - 1	3
1 - 2	2
2 - 3	1
3 - 4	29
4 - 5	3
5 - 6	3

SEX WISE DISTRIBUTION OF DIFFERENT TYPES OF MORPHOEA

Incidence of morphoea was more in females

The female to male ratio was 2 : 1

TABLE - 10

S.No	Clinical types	Female	Male	Total
1	Linear type	11	4	15
2	Plaque type	9	5	14
3	Frontoparietal	4	4	8
4	Gen. Morphoea	2	0	2
5	Mixed	2	0	2
6	Total	28	13	41

SYMPTOMS

The most common presenting complaint was disfigurement - 63%

TABLE - 11

Complaints	No. of patients
DISFIGUREMENT	26
↓ SENSATION	4
PAIN	4
HEAD-ACHE	1
OEDEMA	1
ARTHRITIS	2
SHORTENNING	1
CONTRACTURES	3
HEMIATROPHY	1

ASSOCIATED CONDITIONS

Associated disorders seen were as follows ;

TABLE - 12

Associated features	No. of patients
MUCOSAL LICHEN PLANUS	1
VITILIGO	1
LICHEN SCLEROSUS ATROPHICUS	1
CAFÉ AU LAIT MACULES	1
RHEUMATOID ARTHRITIS	1

NUMBER OF LESIONS SEEN IN MOST PATIENTS

The maximum number of lesion encountered in a patient – 6

The number of patients with single lesions were 33 – (80%)

TABLE -13

No. of lesions	No. of patients
1 LESION	33
2 “	2
3 “	2
4 “	2
5 “	0
6 “	1

SITE COMMONLY INVOLVED

The most common site was lower limbs – 39%

Bilateral involvement of lower limbs was present in 2 patients

The next most commonly affected area was head

TABLE - 14

FACE & SCALP	8
CHEST	2
ABDOMEN	6
BACK	1
UPPER LIMBS	4
LOWER LIMBS	16
BIL. LOWER LIMBS	2

EOSINOPHILIA

Percentage of patients with eosinophilia was 66%

TABLE – 15

Eosinophils > 400/µl	**No. of Patients**
TOTAL NO. OF PATIENTS INVESTIGATED	41
TOTAL NO. OF PATIENTS WITH ↑ EOSINOPHILS	27
PERCENTAGE OF PATIENTS WITH ↑ EIOSINOPHILS	66%

PATIENTS WITH RAISED ESR

Percentage of patients with raised ESR was 41%

TABLE – 16

(ESR > 30mm/hr)	**No. of Patients**
TOTAL NO. OF PATIENTS WITH INVESTIGATED	41
TOTAL NO. OF PATIENTS WITH ↑ ESR	17
PERCENTAGE OF PATIENTS WITH ↑ ESR	41%

HISTOPATHOLOGICAL EXAMINATION

Most of the patients showed feature of late morphoea

Percentage of patients with late morphoea was 80%

TABLE - 17

NO. OF PATIENTS WITH EARLY MORPHOEA	8
NO. OF PATIENTS WITH LATE MORPHOEA	33

ANA POSITIVITY IN DIFFERENT TYPES OF MORPHOEA

(1/10 ++,1/40 ++)

Percentage of patients with ANA positivity was 24%

Percentage of ANA positivity in patients with linear morphoea was 14%

TABLE - 18

S.No.	Different types of morphoea	ANA positivity
1	LINEAR MORPHOEA	6
2	GEN. MORPHOEA	1
3	PLAQUE	2
5	FRONTOPARIETAL	1
	TOTAL	10

RHEUMATOID FACTOR

Rheumatoid factor was positive in one patient – 24 units

C REACTIVE PROTEINS

C reactive proteins was positive in one patient – 320 ng/dl

ANA PATTERN

The most common ANA pattern seen was homogenous type

TABLE - 19

HOMOGENOUS PATTERN	9
SPECKELED PATTERN	1

PRECIPITATING FACTORS

Trauma, organophosphorus compounds and injections have precipitated morphoea in following patients. Trauma (physical and iatrogenic factors) can precipitate morphoea.

TABLE - 20

TRAUMA	2
ORGANOPHOSPHORUS COMPOUNDS	2
INJECTIONS	2

RADIOLOGICAL ABNORMALITIES

Relevant radiological examination of patients with limb shortening, frontoparietal and linear morphoea were undertaken. Radiological examination of spine was carried out in all patients with linear morphoea.

The percentage of linear morphoea with spina bifida occulta was 6%.

TABLE - 21

TOTAL NO. OF PATIENTS WITH LINEAR MORPHOEA SCREENED	15
NO. OF PATIENTS WITH RADIOLOGICAL ABNORMALITY (SPINA BIFIDA OCCULTA)	1
PERCENTAGE OF PATIENTS WITH ABNORMALITY	6%

DISCUSSION

In this study of 41 patients the incidence of morphoea was found to be 1 in 1,000. (Tab.1) Incidence of various clinical types of Morphoea are as follows: (Tab. 2)

Plaque-35%

Linear-36%

Generalaized-5%

Mixed-5%

Frontoparietal-19%

A study by Christianson H.B. of 235 cases showed an incidence of 35% of plaque type, and 49% of linear type and 19% of generalized morphoea.[5]

The youngest person in this study was a 4 year old female child and the most aged person was a 49 year old woman. (Tab.3). In Christianson's study the youngest person was a 1 year old child and the oldest was a 76 year old person. In this study, the maximum number of cases was between 10 to 25 years of age, whereas in Christianson's study, the peak incidence was between 20 to 40 years. In this study the incidence of morphoea below 10 years was 9% and below20 years was 48%. In Christianson's study incidence below 10 years was 1.5% and below 20years was 7.2%.

In this study incidence of plaque type below 10 years was nil and between 20 to 50 years was 34%. (Tab.4). In Christianson's study incidence of the same below 10 years was 10% and between 20 to 50 years was 75 %.

In this study incidence of linear type below 10 years was 2% and below 20 years was 7.2%. (Tab.5). Christianson's study showed 10% incidence below 10 years and 75% below 20 years.

In this study incidence of generalized morphoea between 11 and 50 year were 2% (Tab.6) whereas Christianson's study showed 80 %.

Incidence of frontoparietal morphoea peaked between 10 and 30 years of age. (Tab.7). Maximum incidence of mixed(linear and plaque) type of morphoea was below 10 years of age. (Tab.8).

The average duration of evolution in this study was 2 to 3 years. (Tab.9). In Christianson's study, the average time was 3 to 5 years.

The incidence of morphoea in this study was more in females. The sex ratio between females to males was 2:1. The sex ratio in Christianson's study was 3:1 male. (Tab.10).

The most common complaint of the patients was disfigurement. (63 %) (Tab.11). In Christianson's study 44% of patients presented with pain and arthralgia whereas in this study pain in near by joints was the presenting complaints in only two (5%) of the patients. One of them had hemiatrophy on the right side of the trunk. Two patients had flexion contractures of the joints.

Four persons presented with loss of sensation. There was oedema preceding the appearance of lesion in one patient. There was shortening of 1cm of the lower limbs of one child due to soft tissue contracture. One patient with frontoparietal morphoea complained of head ache and ophthalmologist diagnosed him of having myopia and he was prescribed spectacles. Despite wearing spectacles he continued to have head ache and after which he did not report for follow up. Head ache may be an association of morphoea.[146]

Associated disorders seen in patients in this study includes mucosal lichen planus, vitiligo, rheumatoid arthritis, café au lait macules and lichen sclerosus et atrophicus. (Tab. 12). These associations have also been reported in various other studies. Finklestein E, reported a case of vitiligo with morphoea.[135] Winkleman reported a case of lichen sclerosus with morphoea.[136] Rheumatoid arthritis and lichen planus has also occurred frequently with morphoea.[146]

The maximum number of lesions in one person was six. (Tab.13). Most patients however had only single lesions. (80%)

The most commonly affected site was lower limbs. (39%) (Tab.14). Two of the patients showed bilateral involvement of lower limbs. The next common site was head and third most commonly affected area was the abdomen and chest.

The serological investigations showed eosinophilia and raised ESR. Eosinophilia was present in 66% of patients. (Tab.15). Raised ESR was seen in

41% of patients. (Tab.16) A study conducted by Flagana showed similar findings.[137].

Histopathological examination of biopsy from patients showed features of late morphoea in 33 patients (80%) and early morphoea in the rest (20%) of the patients. (Tab. 17). In late morphoea there was no inflammatory infiltrates and the epidermis was atrophied with loss of rete ridges. The collagen in the dermis was thick, hypertrophied, homogenised, hyalinized and hypocellular. There was also high uptake of eccrine glands. The glands were also atrophic and adipocytes were absent. In early morphoea, inflammatory infiltrates was seen extending up to the eccrine glands and also around perivascular spaces. Endothelial swelling was also seen in blood vessals. Collagen bundles were only slightly thickened.

ANA positivity was 24% in this study.(Tab-18). Signsen et al in his study showed 40% positivity. He also showed that ANA positivity was more in children and in patients with linear morphoea. In this study ANA positivity in linear morphoea was 14%.

Rheumatoid factor was also positive in one patient of morphoea and incidentally he was also having rheumatoid arthritis. He was also positive for C reactive proteins.(320ng/dl). Singsen et al in his study has proved similar association.

Among the 10 patients with ANA positivity, homogenous pattern was seen in 9 patients (90%) one showed speckled pattern of deposits. (Tab.18).

There was history of trauma preceding lesions in 2 patients and there was history of administration of injection(nature unknown) in 2 of patients preceding the appearance of lesions. This shows that trauma could be precipitating factor for development of morphoea as reported in literature.[87]There were 2 farmers in this study with increased exposure to organophosphorus compounds. (Tab. 20). No definite conclusions regarding this factor could be drawn from the above history because of the small sample size.

One child with linear morphoea showed spina bifida occulta on radiological examination of the spine. (6%) (Tab.21) Christianson showed 47% of association of spina bifida occulta with linear morphoea. Rubin et al also showed a similar association.[101]

CONCLUSION

* The incidence of Morphoea in Government General Hospital during the period of September 2004 to September 2006 was 1 in 1000.

* The incidence of various types of Morphoea were as follows

PLAQUE TYPE	34%
LINEAR TYPE	36%
GENERALIZED TYPE	5%
MIXED TYPE	5%
FRONTOPARIETAL TYPE	20%

* The female to male sex ratio was 2:1.

* The maximum number of patients were in the age groups of 10 to 25years of age.

* Linear morphoea was more commonly seen in lower limbs.

* The main complaints of patients was disfigurement.

* One of the precipitating factors was found to be trauma.

* Serological investigations showed eosinophilia in 66% of patients.

* ESR was raised in 41% of patients.

* ANA was positive in 24% of patients.

* Homogenous pattern of ANA was most commonly seen.

* The associated autoimmune disorders seen were lichen planus, vitiligo and rheumatoid arthritis.

* The other associated anomalies seen were spina bifida occulta, café au lait macules, lichen sclerosus atrophicus.

* Histopathological study showed compatibility with late morphoea in most of the patients.

* There was no incidence of morphoea in patient's relatives.

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KEY TO MASTER CHART

F	-	FEMALE
M	-	MALE
Ass	-	Associated
Abd	-	Abdomen
RLL	-	Right upper limb
LLL	-	Left upper limb
LT Head	-	Left Head
RT Head	-	Right Head
SP BOC	-	Spina bifida occulta
½ Body	-	Hemiatrophy
HPE	-	Histopathological examination
HB%	-	Haemoglobin
CRP	-	C reactive proteins
PPPT Factors	-	Precipitating factors
COS	-	Cosmetic problem
Dec Sen	-	Decreased sensation.
Contr	-	Contrature
Loss of hai	-	Loss of Hair
P	-	Plaque type of morphoea
LM	-	Linear type of morphoea
FP	-	Frontoparietal morphoea
GM	-	Generalized morphoea
LSA	-	Lichen sclerosus et atrophicus
LP	-	Lichen Planus

PROFORMA

Case No.:
Complaints:
Duration: Onset: Insidious Rapid
No of lesions:

SITE

Head: Face: Scalp :
Limbs: Upper: Lower:
Trunk: Chest: Buttocks: Back :
Abdomen:

ASSOCIATED COMPLAINTS:

Disfigurement: Contracture: Pain: Arthritis:
Raynaud's phenomenon: Loss of sensation: Decreased sweating:
Loss of hair: Disparity in size of limbs:
Headache: Seizures:

SYSTEMIC COMPLAINTS:

Difficulty in swallowing : Breathlessness:
Dyspnoea : Palpitations:
Past history:
Trauma :
Exanthematous fever:
Pencillamine:
Injections Vitamin K or any other injection
B.C.G Vaccination:
Drugs:
Pencillamine: Bleomysin : Cocaine:
Pentazocine: Bromocriptine : Docetaxel:
Antiepilepticdrugs:

OCCUPATIONAL EXPOSURE:

Vinyl chloride (laundering): benzene (dying):
Toluene: Epoxy resins (film making):
Radiation exposure: Miscellaneous:
Silicon implants:
Exacerbation during pregnancy:
Metabolic disorders like D.M /Phenylketonuria:
Treatment taken:
Family members having similar problems: Relation:

GENERAL EXAMINATION:

Anaemia: Cyanosis: Pedal oedema:
lymphadenopathy:
J.V.P:
Jaundice: Clubbing : Purpuric spots:
Vital signs: P.R: B.P: TEMP: R.R:
Ocular examination:
Lid ptosis: Oedema: Extra ocular muscle weakness:
Enophthalmous: Iris atrophy: Heterochromia iris:
Intraoral examination:
Tongue: Crowding of teeth: Gingival atrophy::

SYSTEMIC EXAMINATION:

C.V.S: R.S: C.N.S: PER/ABD:
Musculoskeletal examination:
kyphosis/scoliosis: Rib anomaly:
Contracted pelvis:
Atrophic clavicle: Disparity of limb length:
Absent pectoralis major: Shortened ulna: Atrophy of face:

DERMATOLOGICAL EXAMINATION:

Single: Multiple Hyper/hypo pigmented plaques:
Hide bound: Well circumscribed: Border:
Loss of hair :
Elevated/depressed/nodular surface : Hypoanesthetic.
Special feature:
Calcinosis/telengectasia/tenderness/oedema
/attachment to underlying Structures:
Hair: Nail: Genitalia: Palms & soles:

INVESTIGATIONS:

T.C: D.C : E.S.R: Blood sugar:
Blood. Urea: Serum Creatinine:
Liver function test:
Urine albumin: Sugar Deposits:
Platelet count: X-Ray: spine: limbs: Skull:
E.C.G:
A.N.A Titre: Rheumatoid Factor: 'C'. Reactive Proteins:
H.P.E: Early/late. Special features:

OPHTHALMOLOGY OPINION:
NEUROLOGY OPINION:

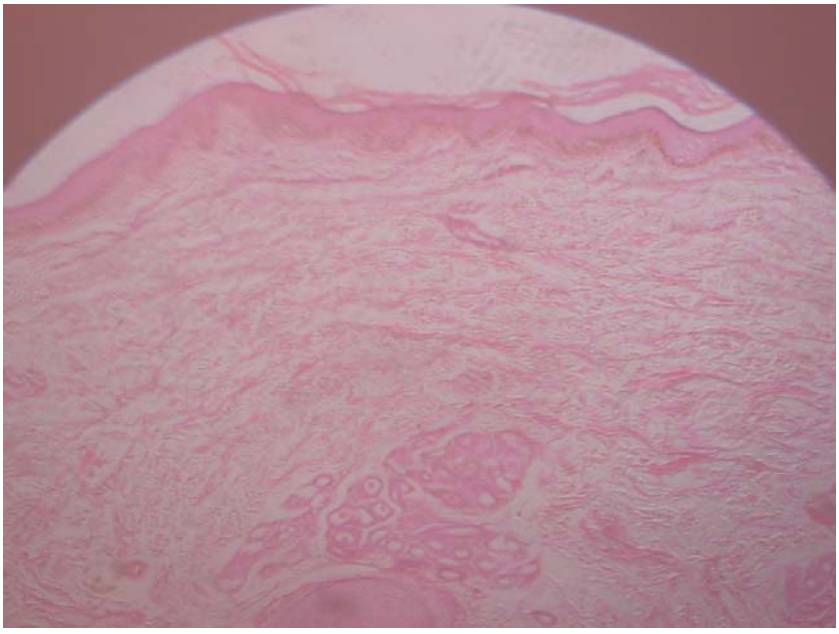
PLAQUE TYPE OF MORPHOEA WITH LILAC COLOURED BORDERS

PLAQUE TYPE OF MORPHOEA WITH HYPERPIGMENTATION

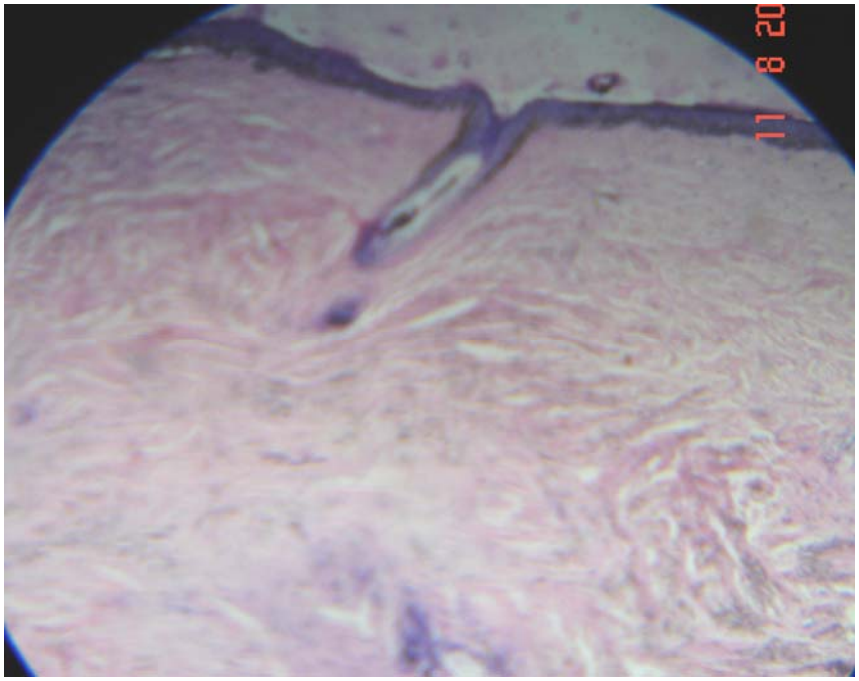
A SMALL BOY WITH FRONTOPARIETAL MORPHOEA SHOWING ALOPECIA

A GROOVE DEVELOPING ON THE FACE IN EARLY FRONTOPARIETAL MORPHOEA

HISTOPATHOLOGY OF LATE MORPHOEA SHOWING HYPERTROPHIED, HOMOGENIZED AND HYLANISED COLLAGEN. THE SWEAT GLANDS ARE ATROPHIC, ADIPOCITES ARE ABSENT AND ARE SORROUNDED BY NEWLY FORMED COLLAGEN



HISTOPATHOLOGY OF EARLY MORPHOEA SHOWING INFLAMMATORY INFILTERATES IN THE DERMIS. THE COLLAGEN IS ALSO OEDEMATOUS



LINEAR MORPHOEA ON THE THIGH

THE SAME PERSON SHOWNG LINEAR MORPHOEA EXTENDING ON TO THE LEGS

THE SAME PERSON SHOWING ATROPHY ON ONE SIDE OF THE CHEST

LINEAR MORPHOEA ON THE LEGS PRESENTING WITH HYPER PIGMENTATION

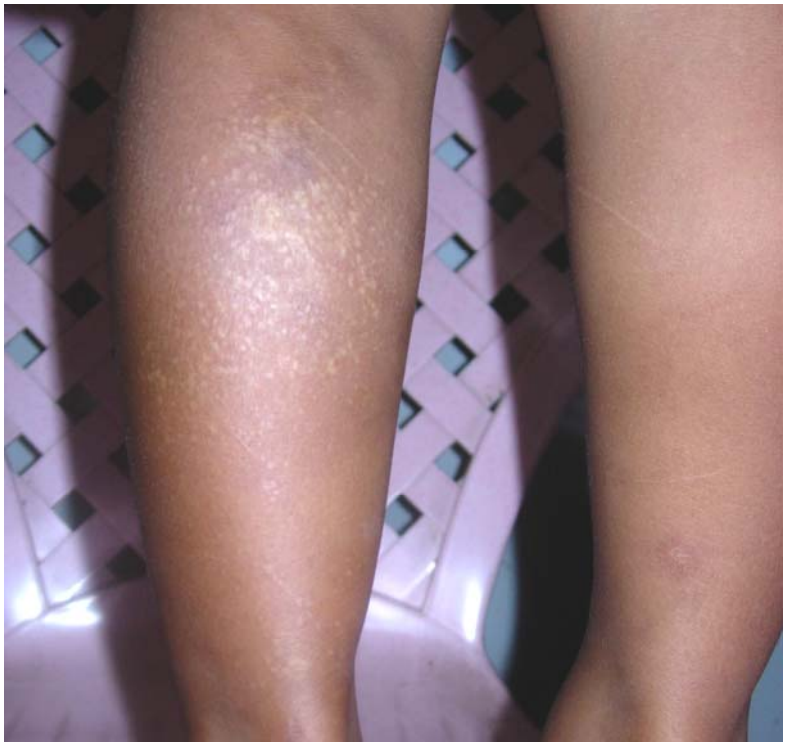
A PATIENT WITH GENERALIZED MORPHOEA SHOWING LESIONS ON THE RIGHT SIDE OF THE CHEST

THE SAME PERSON SHOWING LESIONS EXTENDING ON TO THE GLUTEAL REGION

LINEAR MORPHOEA CROSSING OVER THE WRIST PRODUCING CONTRACTURE

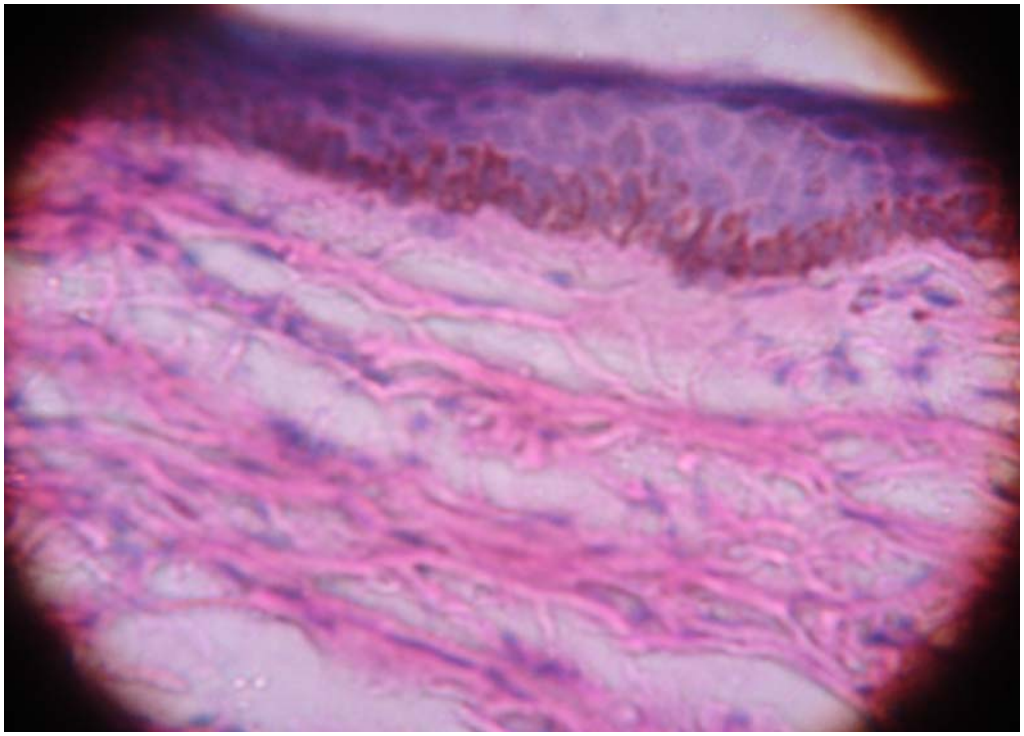
WOMAN WITH GENERALIZED MORPHOEA ALSO HAVING ORAL LICHEN PLANUS

A PATIENT WITH MORPHOEA ALSO HAVING VITILIGO



LICHEN SCLEROSUS EI ATROPHICUS PRESENT IN A PATIENT OF MORPHOEA

HISTOPATHOLOGY SHOWING EARLY MORPHOEA IN HIGHER MAGNIFICATION. PLENTY OF INFLAMMATORY INFILTERATES CAN BE SEEN IN THE DERMIS



X-RAY OF SPINE SHOWING SPINA BIFIDA OCCULTA