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PANSCLEROTIC MORPHOEA WITH PSEUDOAINHUM - A CASE REPORT

Dermatology

KEYWORDS: Panclerotic morphoea, pseudoainhum, low dose methotrexate,

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ABSTRACT

Panclerotic morphoea is a rare and severe disabling form of morphoea with the onset usually in children below the age of 14 years[1]. It is characterized by sclerosis of the skin, subcutis, fascia, muscle and even bone, associated with contracture, deformity, joint stiffness and growth retardation. This form has an aggressive course but spontaneous remission can occur in some cases[2]. We report a case of 9 old year girl with tautness of the skin, involving the right foot and leg with stiffness of the ankle and knee joints and deformity of the right foot, associated with a constriction ring involving the right 3rd toe. Clinical examination revealed a linear, indurated plaque extending from right foot up to upper 3rd of the leg with deformity of right foot and knee and joint stiffness. She also had a pseudoainhum involving the right 3rd toe. The activity of the disease was assesed by histopathological examination, relevant blood examination-eosinophilia, elevated ESR[5], and with radiological examination which showed soft tissue edema and bony involvement. The patient was started on low dose methotrexate. She showed little improvement with treatment. The case is being reported for in view of its rare occurrence.

Introduction:

Panclerotic morphoea is a rare severe mutilating form of morphoea. It is characterized by sclerosis of the dermis, subcutis, fascia, muscle and even bone with the onset in children before the age of 14 years.

Case details:

History:

A 9 year old girl presented with tautness of skin over the right foot and leg associated with stiffness of right ankle joint and difficulty in walking since 2 years. At the onset, 2 years back there was tautness of skin, started initially over the dorsum of right foot with gradual extension up to upper third of the right leg associated with deformity of the right foot, and knee. There was no prior history of trauma, surgery, injection or drug intake. She had no history of Raynaud's phenomenon or symptoms suggestive of systemic involvement.

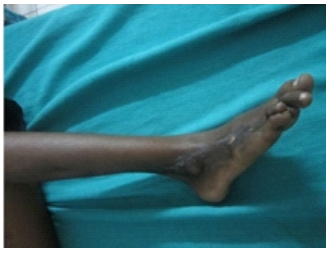

Fig 1 : Shows tautness of skin over the right foot and leg

Examination:

General examination was normal.

Dermatological examination revealed a linear, hyperpigmented, indurated, atrophic depressed plaque extending from right 3rd toe along the lateral 3rd of the dorsal foot to upper 3rd of the posterior aspect of leg. The plaque was dry and hypoaesthetic, and it was attached to the underlying deeper structures. The length and girth of the right leg were decreased by 5 centimeters and 3 centimeters respectively when compared to opposite side. There was restriction of all movements of the right ankle and extension at knee joint was restricted. Flexion deformity of right knee, talipus and valgus deformity of right foot were present. Pseudoainhum in right 3rd toe was noticed, and overriding of toes was present.

Investigations:

Blood investigations revealed elevated ESR and eosinophilia. Anti nuclear antibody was negative.

Histopathological examination revealed normal epidermis,and dermis showed interstitial, perivascular and periappendageal lymphocytic infiltrates, and thickened homogenous hypocellular eosinophilic collagen fibers in dermis and subcutis. Atrophy of eccrine glands and its high uptake were present. Thickening of dermal blood vessel walls were seen.

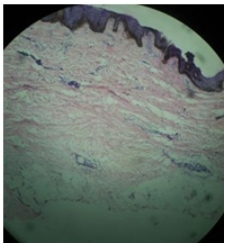

Fig 2:

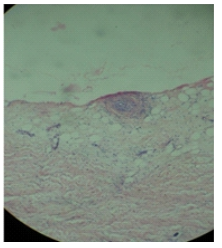

Fig 3:

Fig 2&3: Shows thickened homogenous hypocellular eosinophilic collagen fibers in dermis and subcutis

MRI examination of the right leg revealed soft tissue edema, fascial thickening and increased enhancement in calcaneum.

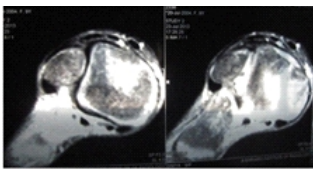

Fig 4

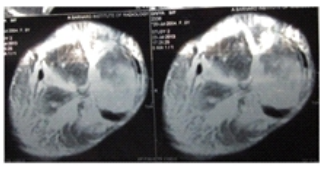

Fig 5

Fig 4 &5: Shows soft tissue edema, fascial thickening and increased enhancement in calcaneum.

Patient was started on methotrexate 2.5mg / wk and, advised physiotherapy and plastic surgical correction for the contracture and deformity.

Discussion:

Pansclerotic morphoea is a rare severe mutilating form of morphoea involving dermis, subcutis, muscles and even bones, usually affecting the children below 14 years of age.[1,2] It is an immunologically mediated condition causes endothelial cell damage, inflammation and release of cytokines, the platelet derived growth factor(PDGF), transforming growth factor-beta(TGF) which stimulates the fibroblast to produce increased glycosaminoglycan, fibronectin and collagen, and decreased extracellular matrix resulting in sclerosis[2,3]. The triggering factors include trauma, surgery, injection, vaccination, infections like Borreliosis, measles and varicella, and drugs.

It is clinically characterized by an asymmetrical, linear, smooth, shiny, sclerotic plaque usually affecting the trunk and extremities, scalp and face with sparing of finger tips and toes. The plaque is dry, hypoaesthetic and attached to the deeper tissues. Involvement of the deeper structures causes contracture, deformity and stiffness of the joint[5]. Intense pain may occur due to cutaneous nerve involvement. Raynaud's phenomenon and systemic involvement are usually absent[2].

Complications include contracture, deformity, joint stiffness, growth retardation, pseudoainhum and squamous cell carcinoma[11] from long standing ulcers. Associations like spinal abnormalities, migraine, abdominal pain and dermatological conditions like vitiligo, generalized icthyosis, pigmented and vascular naevi have been reported. This condition should be differentiated from melarheostosis[6].

On histopathological examination, in the early inflammatory stage, the epidermis is normal and the reticular dermis shows interstitial lymphoplasmacytic infiltrates among slightly thickened collagen bundles and mild vascular changes like endothelial swelling, edema of the vessel wall. The late sclerotic stage is characterized by disappearance of the inflammatory infiltrate,and presence of thickened, homogenous, hypocellular, hyalinized and hypereosinophilic collagen in dermis and subcutis. Eccrine glands are markedly atrophic, and they seem to lie higher in the dermis due to replacement of subcutaneous fat by newly formed collagen fibers[4]. Fascia shows fibrosis and sclerosis. Muscle fibers appear vacuolated. Other laboratory findings include raised ESR, eosinophilia, hypergammaglobulinemia, ANA positivity in 40% and, positive Anti ssDNA in 53.3% cases. Eosinophilia, Anti ssDNA, and IgM Rheumatoid factor are useful indicators of disease activity. Patients with Anti ku antibody carry poor prognosis and are prone for systemic sclerosis. MRI and Ultrasonogram can be done to assess the depth of the disease involvement.

Treatment with PUVA or low dose UVA1, low dose methotrexate[7] with steroid pulse therapy will be beneficial in the early active disease. Other options include d-penicillamine, phenytoin, griseofulvin, chloroquine, acitretin, ciclosporin, mycophenolate mofetil and bosentan. Recent therapeutic modalities like imatinib mesylate[9], an N-demethylated piperazine derivative and Autologous Stem Cell Transplantation[8] can be used with good results. Late sclerotic stage has to be managed with physiotherapy and surgical correction for contracture and deformity[10].

Our patient had onset at 7 years of age and diagnosis of pansclerotic morphoea was made based on clinical examination, aided by skin biopsy. Histopathological findings with inflammatory infiltrate, and blood eosinophilia were suggestive of active disease, hence the patient was started on low dose methotrexate at 2.5 mg/week.

Conclusion:

This case is being presented to highlight its rare occurrence with the rare complication of pseudoainhum.

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