Unilateral linear pansclerotic morphea affecting face and limbs

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ABSTRACT

Disabling pansclerotic morphea is a rare atrophying and sclerosing disorder of the subcutaneous tissue, muscle and bone. It is characterized by atrophy of the skin, subcutaneous fat, muscle and bone involving half of the face. In some patients the atrophic lesions extend to involve the ipsilateral or contralateral upper and lower limbs with radiological evidence of hemiatrophy. The patients may present with arthralgia, convulsions or cramps. We report a case of a woman with deformity of face, and left upper and lower limbs that had started as an indurated plaque on the left half of forehead at the age of 5 years and had gradually enlarged, followed by the development of atrophic changes in left eye. The case is being reported in view of its rare occurrence.

Key Words: Disabling pansclerotic morphea, Parry Romberg syndrome, Morphea, En coup de sabre

INTRODUCTION

Disabling pansclerotic morphea is a rare atrophying and sclerosing disorder of the subcutaneous tissue, muscle and bone. This disease is characterized by atrophy of the skin, subcutaneous fat, muscle and bone involving half of the face. Generalized full thickness involvement of trunk, extremities, face and scalp is described in disabling pansclerotic morphea. We report here one such case with widespread unilateral affection.

CASE REPORT

A 20-year-old woman presented with deformity of the face, and left upper and lower limbs from 5 years of age. It had started as an indurated plaque on the left half of the forehead and had gradually enlarged. This was gradually followed by the development of atrophic changes in the left eye, facial deformity and left upper and lower limbs. There was no history of photosensitivity or symptoms suggestive of Raynaud’s phenomenon. She did not have dysphagia or cough. There was no history of trauma. Her family history was not contributory. Systemic examination did not reveal any abnormality clinically, other than the limitation of movements due to contractures.

Physical examination revealed marked facial asymmetry. A linear, paramedian, atrophic, depressed, indurated skin colored plaque was present on the left side of the forehead, extending up to the coronal suture. [Figure
The left half of the nose was atrophic. There was shortening of the left upper limb, a linear atrophic indurated plaque extending to the shoulder, and a flexion contracture at the left elbow [Figure 2]. Flexion deformity and contractures were also seen in the left lower limb at the knee and ankle [Figure 3]. The right arm and forearm were 7.5 cms and 10 cms longer respectively, compared to the left side. The right leg was 2.5 cms longer than the left, when measured from patella to medial malleolus.

Radiological examination revealed soft tissue atrophy in the lower third of the left thigh and leg, and periosteal reaction in the left fibula. The left greater and lesser trochanters were small in size. An X-ray chest showed right basal consolidation. AP and oblique views of the left foot showed evidence of sclerodactyly, while the first and second metatarsals revealed gigantism.

A skin biopsy from a plaque on the forehead and on the forearm showed flattening of rete ridges, extensive fibrosis and sclerosis in the dermis and subcutaneous tissue. Due to lack of facilities, serological tests for *B. burgdorferi* infection and ANA study could not be performed. Other hematological parameters were within normal limits.

The patient was diagnosed as unilateral pansclerotic morphea. She was advised physiotherapy but was lost for follow-up, after initial physiotherapy failed to produce substantial improvement.

DISCUSSION

Localized scleroderma/morphea is a benign well-circumscribed lesion in the skin. It is characterized by round, oval or irregular plaques indurated and bound down to the underlying tissues and rarely accompanied by atrophy of the underlying structures.

Peterson et al developed a revised classification of localized scleroderma [Table 1]. Inclusion of lichen sclerosus et atrophicus into the group of morphea is controversial but transitions between morphea and lichen sclerosis et atrophicus as well as coexistence of both are described. It is unclear whether the heterogeneous clinical picture of morphea represents one disease with varying morphology or a summation of different diseases.

The morphological expression of localized scleroderma thus forms a spectrum with lichen sclerosus et atrophicus at one end and disabling pansclerotic morphea at the other end, and encompassing the other clinical variants as shown in Table 1.

Pansclerotic disabling morphea usually starts before the age of 14 years while our patient first developed lesions...
Table 1: Proposed classification of morphea

<table>
<thead>
<tr>
<th>Classification</th>
<th>Subclassifications</th>
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<tbody>
<tr>
<td>a. Plaque Morphea</td>
<td>Lichen sclerosus et atrophicus Morphea en plaque Guttate morphea Atrophoderma of Pasini and Pierini Keloidal /nodular morphea</td>
</tr>
<tr>
<td>b. Generalized Morphea</td>
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<td>c. Bullous Morphea</td>
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<td>d. Linear Morphea</td>
<td>Linear scleroderma En coup de sabre associated with progressive hemifacial atrophy</td>
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<tr>
<td>e. Deep Morphea</td>
<td>Subcutaneous morphea Eosinophilic fascitis Morphea profunda Disabling pansclerotic morphea of children</td>
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at the age of 5 years. A marked female preponderance is observed at a 3:1 ratio.[6] Ipsilateral atrophy of the limbs with radiological evidence of bony affection was observed along with a lesion of the face in our patient. A case of morphea with polyarthritis and atrophy of the limbs on opposite side of the body was reported in a 28-year-old woman.[7] However, our patient did not have arthritis, cramps or convulsions as reported by other authors.[7-8]

The etiology of this condition is still not known. Some patients with morphea, particularly in Europe, have sclerosis due to Borrelia burgdorferi infection. In such cases, if the lesions are not too sclerotic, they can disappear with prolonged courses of oral antibiotics. Morphea can follow X-ray irradiation of breast cancer.[9]

Appropriate serological testing to rule out B. burgdorferi infection is advised. On biopsy epidermis may appear normal or show atrophic changes. Pansclerotic lesions show fibrosis and disappearance of the subcutaneous tissue. Silver stains on skin biopsies may be performed to rule out B. burgdorferi infection.

Diagnosis is essentially by clinical examination aided by skin biopsy. The disease is slowly progressive and spontaneous remission can occur. There is no effective treatment. In patients with early involvement beneficial effect in the form of reversal of sclerosis with high dose of parenteral penicillin or ceftriaxone given in several courses over a time span of several months is reported.[9] The best response is achieved when it is combined with oral corticosteroids.[9]

Phototherapy with UVA-1 (340-400 nm) has been found to be effective. The mechanism of action is thought to be by the induction of interstitial collagenase [matrix metalloproteinase-1 (MMP-1)]. Collagenase initiates degradation of types I and III collagen and plays a key role in the remodeling of dermal collagen. UVA-1 phototherapy can also induce a variety of cytokines and soluble factors, resulting in immunomodulation. However, some authors opine that UVA-1 phototherapy is not very easy or successful because of prolonged irradiation times and the disfiguring hyperpigmentation of the irradiated areas.[9]

Other therapeutic modalities tried with varying results are calcitriol, D-penicillamine, low dose methotrexate, topical calcipotriene, diphenyl hydantoin and antimalarials. Progressive facial hemiatrophy may be amenable to bone grafting, tissue expansion and microsurgical reconstruction.[9]

REFERENCES