Extensive morphea profunda with autoantibodies and benign tumors: A rare case report

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ABSTRACT

The term deep morphea describes a variant of morphea (localised scleroderma) in which inflammation and sclerosis are found in the deep dermis, panniculus, fascia or superficial muscle. It is sometimes associated with autoantibodies. We report the case of a 49 year-old male who had morphea profunda (radiologically and histopathologically confirmed) affecting mainly the left side of the body with face, trunk and limb involvement, along with autoantibody production and associated neurofibromas and lipomas.

Key words: Autoantibodies, benign tumors, morphea profunda

INTRODUCTION

The term scleroderma describes a disease of unknown etiology that causes fibrosis of skin and internal organs. Morphea differs from systemic sclerosis by the absence of clinically detectable systemic involvement.[1] Histologically, all subtypes show homogenisation of collagen bundles while the distinguishing features are the distribution and depth of skin involvement.[2]

CASE REPORT

History and Examination

A 49-year-old male presented to the Neurology department with complaints of radiating pain along the left lower limb. He was referred to the Dermatology department in view of hyperpigmented lesions over the back, abdomen and limbs. The patient first noticed these skin lesions at the age of 15 years when they were present over the face. Later, these lesions spread to gradually involve the abdomen, back, upper and lower limbs over a period of 20 years, i.e. till 35 years of age. The lesions were completely asymptomatic and have been static since then. He however did not consult any doctor as he did not have any problem due to these lesions. He developed radiating pain along the left lower limb since the past 1 year, which increased in intensity over the past 1 month. He did not have any other associated complaints. The patient denied any history of trauma or drug intake preceding the occurrence of lesions. There was no history of joint pains, cold intolerance and no difficulty in swallowing or breathing. There was also no history of any skin tumors or decreased hearing. There was no history of similar complaints in the family.

On cutaneous examination, he had hyperpigmented atrophic patches over the left side of the face [hemiatrophy, Figure 1]. Central forehead showed a soft swelling with a tendency to slip on palpation, suggestive of a lipoma. Multiple hyperpigmented atrophic patches were seen over the upper back, both upper limbs, left inframammary area, abdomen and left lower limb [Figures 2 and 3]. These patches were indurated and difficult to pinch. There were no skeletal deformities. The patient did not have any axillary or palmar freckling and no café-au-lait macules. Ophthalmic examination revealed normal vision with no Lisch nodules. Hearing was normal. Major systems were normal on clinical examination.

Investigations

Complete blood count, liver function tests, renal function tests and erythrocyte sedimentation rate (ESR) were within normal limits. C reactive protein (CRP) was elevated. Antinuclear antibodies, antihistone and anti double stranded-DNA (anti ds-DNA) antibodies were negative. Rheumatoid factor was strongly positive (122 IU/ml). Urine examination was normal. Chest radiograph was normal.

A deep skin biopsy [Figure 4] taken from the back revealed atrophic epidermis and increased...

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collagenisation in the papillary dermis. There was increased fibrosis around the adnexal structures and subcutaneous fat. Mononuclear inflammatory infiltrate was also seen.

Radiologic investigations
Computed tomography (CT) scan and magnetic resonance imaging (MRI) of the spine done for the radiating pain revealed two tumors at the junction of L₁-L₂ and L₂-L₃ vertebrae suggestive of neurofibromas Both had intra- and extraspinal parts. CT scan and MRI of the brain were normal except for two lipomas in the frontal region. CT scan of the thorax and abdomen were normal, except for loss of subcutaneous fat.

A final diagnosis of morphea profunda with autoantibodies and benign tumors was thus made.

Management
The patient was started on anti-inflammatory drugs for his symptoms. He was then operated for his spinal lesions. The tumors were confirmed to be extradural neurofibromas with intra- and extraspinal components. Laminectomy was done and the tumors were removed. The patient’s limb pain disappeared completely after surgery. As the skin lesions were asymptomatic and static, the patient refused any treatment for them. He also shifted residence shortly after the surgery and was hence not available for regular follow up. He was therefore not given any treatment for skin lesions.

DISCUSSION
The term scleroderma is applied to a spectrum of disorders characterised by thickening of skin and subcutaneous tissue. Two clinical categories can be identified: systemic sclerosis in which visceral lesions are present and morphea in which lesions are limited to the skin.[2] Peterson and co-workers have proposed a classification system for morphea.[3]

Deep morphea syndromes are characterised by the involvement of the deep dermis, subcutaneous tissue fascia or superficial muscle.[3] Eosinophilic fasciitis is a distinct syndrome described by Shulman, consisting of sclerotic fasciitis, peripheral eosinophilia, elevated ESR and hypergammaglobulinemia.[4]

The term morphea profunda was proposed by Person and Su in 1981 to describe generalised inflammatory sclerosis of the panniculus or fascia. The term solitary morphea profunda was used by Whittaker to describe a solitary fibrotic plaque on the upper trunk with histological findings of a dense, mononuclear cell infiltrate mainly in the subcutis, with marked sclerosis and hyalinization of the connective tissue. Morphea profunda is a disorder of unknown etiology but has been described to occur following vaccination[7] and in association with *Borrelia burgdorferi* infection.[8] It is more common in females. Our patient was male. It presents with one or more hyperpigmented, shiny, smooth atrophic patches with the skin bound down to the underlying structures. The sclerosis is not involved with Raynaud’s phenomenon or ulceration of the digits. Our patient did not have any of these. Systemic involvement in the form of abnormal pulmonary function, oesophageal motility changes may be seen. However, our patient did not have any evidence of systemic involvement. He had associated symptomatic spinal neurofibromas on the same side as the predominant skin lesions. Symptomatic spinal neurofibromas, although rare, have been reported, although not in association with morphea. The neurofibromas were not associated with any other manifestation of neurofibromatosis.
The patient also had lipomas, which have not been reported in association with morphea profunda.

Investigations in cases of morphea profunda may show peripheral eosinophilia, high gammaglobulinemia, raised ESR and serologic abnormalities like antinuclear antibodies, anti double stranded-DNA, anti single stranded-DNA, antihistone antibodies and rheumatoid factor. Our patient was rheumatoid factor positive along with a raised CRP, suggestive of an inflammatory process.

Parry Romberg syndrome also presents with hemifacial atrophy and is considered by some to be a variant of morphea. This can also be considered as a differential in our case. The ongoing debate between the partial overlap of these two conditions precludes a clear distinction between them. However, our patient had lesions on the trunk and limbs, did not have any central nervous system manifestations or brain changes and was positive for rheumatoid factor, which is seen more commonly in morphea. Hence, the diagnosis tilts more toward the diagnosis of morphea profunda.

The case is reported in view of extensive morphea profunda without systemic involvement, occurring in a male, with raised rheumatoid factor levels, with associated benign tumours, i.e. neurofibromas (symptomatic) and lipomas and for its rarity.

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