SEVERE GENERALIZED MORPHEA: A DEVASTATING EVENTUALITY IN THE ELDERLY

ABSTRACT

Morphea is a localized form of scleroderma characterized by sclerotic skin plaques. It is an uncommon fibrotic reaction limited to the skin and adjacent structures which is unaccompanied by visceral involvement. Although its cause is unknown; genetic, infectious and autoimmune mechanisms have been suggested in morphea. It is more common among children and young women. Topical corticosteroids, systemic corticosteroids methotrexate, penicillamin and topical tacrolimus 0.1% are used in treatment. UVA1 (340-450 nm) phototherapy may also be helpful. In this report we present a 72 year old man with severe generalized morphea. Besides dwelling on its etiopathogenesis, we are also touching upon the impact of the disease on this geriatric patient’s functional and psychosocial capabilities.

Key Words: Geriatric; Generalized morphea; Joint contractures.

ÖZ


Anahtar Sözcüklər: Geriatrik; Jeneralize morphea; Eklem kontraktörü.
MORPHEA

INTRODUCTION

Morphea is a localized form of scleroderma characterized by sclerotic skin plaques. It is an uncommon fibrotic reaction limited to the skin and adjacent structures which is usually unaccompanied by visceral involvement. It is more common among children and young women and the exact underlying etiology is not yet ascertained. In this case report presenting an elderly male with severe generalized morphea, we touched upon its etiopathogenesis and psychosocial burden.

CASE

A 72 year old man was seen for his complaints of dryness and itching on his back along with alopecia, joint pain and weight loss. He declared that he had lost 20 kg in the preceding 3-4 months. He had a history of syphilis infection 30 years ago and left nephrectomy. His physical examination revealed androgenic alopecia; facial seborrheic keratosis and lentigo. An 8x10 cm hyperpigmented and atrophic lesion was detected in the interscapular region while hypopigmented atrophic lesions were seen in his postauricular area and on his thighs bilaterally. Laboratory analysis—including the tumor markers—was unremarkable except for decreased hemoglobin (12.4 g/dl). The abdominal ultrasonography (USG) and abdominal and thoracic computed tomographies (CT) were normal. Electrodagnostic studies yielded generalized axonal polyneuropathy. With the likely diagnosis of morphea, he was discharged to complete a regimen of fluocortolone (10 mg/day), diclofenac, omeprazol and was called for a control visit.

Six months later, he was admitted to our internal medicine ward with a significant increase in his complaints—thickening of his skin all over the body, generalized itching, joint stiffness and weight loss. The physical examination was consistent with severe scarred alopecia, generalized indurated plaques firm on palpation, widespread hypo- and hyperpigmented areas, limited joint motions especially causing flexion contractures in elbow, knee and ankle joints (Figure 1-3). Consequently, his gait was impaired and he could not walk without assistance. Sensory examination was relevant with generalized hypoesthesia. The laboratory analysis revealed decreased hemoglobin (11.2 mg/dl); increased erythrocyte sedimentation rate (80 mm/h). Anti-nuclear, anti-ds DNA, anti-mitochondrial, anti-microsomal, anti-Scl 70, anti-HIV antibodies were all negative. Serum immunoglobulin and complement levels were normal. VDRL test was positive but...
TPHA was negative. Serum protein electrophoresis was also normal. Control USG and CT evaluations were noncontributory. Esophageal motility, pulmonary function and lung diffusion tests were all normal. The skin biopsy uncovered irregular and thickened collagen bundles with decreased skin attachments which were typically indicative of late stage morphea. We started a combination of salazopyrine, colchisine, steroid and a nonsteroidal anti-inflammatory drug – along with home exercises. Two months later, although the skin findings did not regress, the joint symptoms subsided a little. On his follow up 1 year later, he was found to have significant deterioration - severe joint contractures and generalized obtrusive skin lesions. He was emotionally unstable and declined hospitalization. We offered psychiatric support but he never applied to our clinic thereafter.

**DISCUSSION**

Localized scleroderma is classified into two major groups: morphea and linear scleroderma. Morphea can be classified either as an isolated 1-15 cm plaque lesion or as a generalized form with multiple lesions, according to the clinical presentation and depth of tissue involvement (1). Linear or deep morphea lesions can cause restricted mobility, contractures, and deformity. Central nervous system abnormalities related to craniofacial linear morphea cause muscle weakness. Deep morphea lesions can cause peripheral nerve involvement causing extremity weakness and carpal tunnel syndrome. Ptsosis, extraocular muscle dysfunction, anterior uveitis, episcleritis, glaucoma, xerophthalmia, and keratitis are manifestations of ocular involvement. Craniofacial morphea may show altered dentition, malocclusion, and asymmetry of the tongue besides alopecia, loss of eyebrows and eyelashes (2).

Though its cause is unknown; radiation therapy and infec-

tious, genetic, and autoimmune mechanisms have been sug-
gested in morphea (3-7). However, when generalized morphe-
a is observed atypically in the elderly, one must consider so-
me other malignant disorders that should be ruled out. In
the pertinent literature very few cases are reported — a 50 year old female with a biliary cirrhosis (8), and a case of adult acral cutaneous myofibroma (9). In our patient, all of our diagnostic interventions failed to unmask any concomitant malignancy. The literature focuses on *Borrelia burgdorferi* as a possible etiologic agent for morphea (10,11). Morphea is usually diagnosed by clinical examination. The diagnosis is sometimes confirmed with blood tests, skin biopsies, or other methods. Antinuclear antibodies, antihistone antibodies, and rheumatoid factor may be present. Furthermore, antibodies to single-stranded DNA (ssDNA) are seen in over 50% of generalized morphea cases.

In addition to all these etiological controversies, we also highlight the psychosocial impact of this clinical entity on the quality of life of an elderly male. Beyond the salient skin lesions and the various complaints pertaining to them, joint problems were also disturbing the patient significantly. We considered the joint contractures to stem from the skin stiffness nearby, surely not from an arthritic process nor from his generalized axonal polyneuropathy. Though very rare, neuropathies in these patients usually ensue due to either vasculitis (axonal) or enwrapping collagen fibrils (demyelinating) (12,13). Anyhow, the gait of the patient was impaired, which indeed handicapped him. He could not walk or could walk with difficulty even with assistance. Unfortunately, he was brought to the hospital with a stretcher on his last visit. There was an obvious psychosocial burden reducing the quality of his elderly years while his peers were ‘growing roses’.

Generally plaque-type morphea often undergoes spontaneous resolution over a 3- to 5-year period and active lesions can be treated with topical corticosteroids which may reduce inflammation and prevent progression. Systemic corticosteroids alone and/or with methotrexate can be used in treatment of patients with potentially disabling generalized, linear, or deep morphea (14). Penicillamine has been reported as beneficial in small series; but its renal toxicity should be kept in mind (15). In a study, topical tacrolimus 0.1% was found to be effective in active plaque morphea (16). Medium-dose UVA1 therapy due to deep penetration into the dermis is found to be effective in the treatment of localized morphea (2,17).

Overall, our case is exemplifying an untoward clinical sce-
nario in an old man and also reveals the likelihood of a severe course in elderly patients, contrary to how it generally pro-
ceeds in the young — without any disability. Thus, all aspects of morphea should be treated in the elderly.
REFERENCES


