

Yusuf AYDIN¹
İhsan ÜSTÜN²
Elif ÖNDER¹
Adem GÜNGÖR¹



CASE REPORT

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.



OLGU SUNUMU

YAŞLILIKTA YIKICI BİR SONUCA SEBEP OLAN AĞIR JENERALİZE MORFEA

Öz

Morphea sklerodermanın sklerotik cilt plakları ile karakterize olan lokalize bir formudur. Cilt ile sınırlı nadir görülen bir fibrotik reaksiyondur ve viseral tutulum pek görülmemektedir. Nedeni tam olarak bilinmemekle birlikte; genetik, enfeksiyöz ve otoimmün mekanizmalar öne sürülmüştür. Genellikle çocuklarda ve genç kadınlarda görülmektedir. Genellikle klinik seyri iyi olan ve spontan iyileşebilen bir hastalık olmasına rağmen, bazen belirgin morbiditeye de neden olabilmektedir. Bugüne kadar çeşitli ilaçlar denenmiş ve değişik sonuçlar elde edilmekle birlikte hastalığın tedavisinde topikal kortikosteroidler, sistemik kortikosteroidler, methotrexate, penicillamin, topikal tacrolimus 0.1% kullanılmaktadır. UVA1 fototerapisi ile morfea lezyonlarında düzelme görülmektedir. Jeneralize morfea olguları yaşlılarda çok nadir olarak bildirilmektedir. Vakamız eklemelerde hareket kısıtlılığına yol açan kontraktürleri bulunan ve ağır yaygın morfeası olan 72 yaşında bir geriatric erkek hastadır. Hastalığın etyopatogenezi ve geriatric hastadaki fonksiyonel ve psikolojik etkilerine değinilmiştir.

Anahtar Sözcükler: Geriatric; Jeneralize morfea; Eklem kontraktürü.

İletişim (Correspondance)

Yusuf AYDIN
Düzce Üniversitesi Tıp Fakültesi
İç Hastalıkları Anabilim Dalı DÜZCE
Tlf: 0324 336 39 50
e-posta: dryusufaydin@yahoo.com

Geliş Tarihi: 26/04/2009
(Received)

Kabul Tarihi: 18/06/2009
(Accepted)

¹ Düzce Üniversitesi Tıp Fakültesi İç Hastalıkları
Anabilim Dalı DÜZCE
² Mersin Devlet Hastanesi Endokrinoloji MERSİN



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

8×10 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. Electrodiagnostic 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

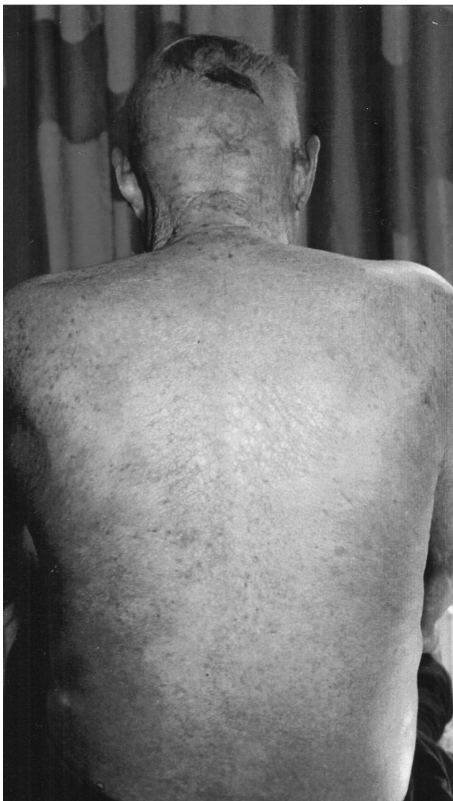


Figure 1— Widespread skin lesions on the back and the scalp of the patient.

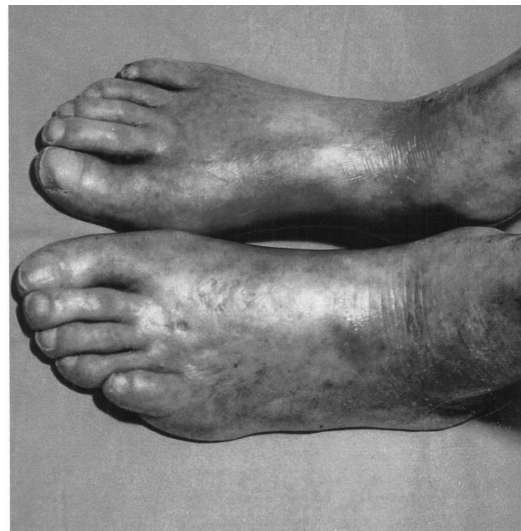


Figure 2— Firm and indurated plaques with hypo- and hyperpigmented areas on the feet.



Figure 3— The flexion contractures of the knee joints with hypopigmented lesions of the cruris.

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, colchicine, 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: Lmorphea 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. Ptosis, 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 suggested in morphea (3-7). However, when generalized morphea is observed atypically in the elderly, one must consider some 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 scenario in an old man and also reveals the likelihood of a severe course in elderly patients, contrary to how it generally proceeds in the young –without any disability. Thus, all aspects of morphea should be treated in the elderly.



REFERENCES

1. Wigley FM. Systemic sclerosis and related syndromes: clinical findings. In: Primer on the rheumatic diseases (Klippel JH, ed), 11th edn. Atlanta, Georgia: Arthritis foundation. 1997, pp 267-72.
2. Bergstrom KG, Schaffer JV. Morphea. Emedicine. [http://emedicine.medscape.com / article / 1065782-overview](http://emedicine.medscape.com/article/1065782-overview). Erişim: 3 Ağustos 2006.
2. Hofmann H, Meyer-Konig U. [Serodiagnosis in dermatological diseases in *Borrelia burgdorferi* infections]. *Hautarzt* 1990;41:424-31.
3. Freeman WE, Leshner JL, Smith JG Jr. Connective tissue disease associated with sclerodermoid features. Early abortion, and circulating anticoagulant. *J Am Acad Dermatol* 1988;19:932-7.
4. Tuffanelli DL. Localized scleroderma. *Semin Cutan Med Surg* 1998;17:27-33.
5. Nagai Y, Hattori T, Ishikawa O. Unilateral generalized morphea in childhood. *J Dermatol* 2002;29:435-8.
6. King DF, Dore RK, Gilbert DJ, Gurevitch AW, Hirose FM. Generalized morphea with peripheral eosinophilia, fasciitis and myositis. *Int J Dermatol* 1980;19:149-53.
7. Suyama Y, Murawaki Y, Horie Y, Hirayama C, Kanbe N, Takenaka M, Shimao S, Ohta G. A case of primary biliary cirrhosis associated with generalized morphea. *Hepato-gastroenterology* 1986;33:199-200.
8. English JC 3rd, Derdeyn AS, Smith PD, Patterson JW. Adult acral cutaneous myofibromas in a patient with generalized morphea. *J Am Acad Dermatol* 2002;46:953-56.
9. Nakashima T, Maeda M, Hayashi T, Kitamura K. A case of generalized morphea with a high titer of anti-*Borrelia burgdorferi* antibodies. *J Dermatol* 1999;26:821-24.
10. Prinz JC, Kutasi Z, Weisenseel P, Póttó L, Battyáni Z, Ruzicka T. "Borrelia-associated early-onset morphea": a particular type of scleroderma in childhood and adolescence with high titer antinuclear antibodies? Results of a cohort analysis and presentation of three cases. *J Am Acad Dermatol* 2009;60(2):248-55.
11. Morita A, Tsuji T. Necrotizing vasculitis in a patient with generalized morphea. *J Am Acad Dermatol* 2001;45:S215-7.
12. Badakov S. Ultrastructural changes of cutaneous nerves in scleroderma. *Folia Med (Plovdiv)* 1992;34:57-64.
13. Kreuter A, Gambichler T, Breuckmann F, et al. Pulsed high-dose corticosteroids combined with low-dose methotrexate in severe localized scleroderma. *Arch Dermatol* 2005;141(7):847-52. Erratum in: *Arch Dermatol* 2005 Sep;141(9):1091
14. Falanga V, Medsger TA Jr. D-penicillamine in the treatment of localized scleroderma. *Arch Dermatol* 1990;126(5):609-12.
15. Kroft EB, Groeneveld TJ, Seyger MM, de Jong EM. Efficacy of topical tacrolimus 0.1% in active plaque morphea: randomized, double-blind, emollient-controlled pilot study. *Am J Clin Dermatol* 2009;10(3):181-7
16. Camacho NR, Sánchez JE, Martin RF, González JR, Sánchez JL. Medium-dose UVA1 phototherapy in localized scleroderma and its effect in CD34-positive dendritic cells. *J Am Acad Dermatol* 2001;45(5):697-9.