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A CASE OF LOCALIZED SCLERODERMA (MORPHEA) SUSPECTED OF MALIGNANT MELANOMA

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ABSTRACT

We present a case of morphea suspected of malignant melanoma, which was resistant to topical corticosteroid, but responded well to daily 5mg of prednisolone. Diagnosis of morphea in the early stage is sometimes difficult, but the presence of lilac ring and dermoscopic finding are helpful. Early diagnosis and systemic therapy are indicated for better prognosis of morphea.

INTRODUCTION

Localized scleroderma (morphea) develops as or multiple sclerotic lesions distributed asymmetrically without systemic organ involvement. Skin lesion starts as a red violaceous fleck and advance to an elastic hard plaque with glossy appearance. Generally morphea lacks characteristic skin manifestations of systemic scleroderma (SSc) including Raynaud phenomenon and disease specific auto antibodies. The prognosis of morphea is good, but some cases with bone involvement leave irreversible deformities. Especially, cases developed in childhood occasionally develop imbalance of extremities and asymmetry of the face. Morphea lesions in the early stage are sometimes difficult to diagnosis because of its variety of clinical manifestation, but respond well to therapy. The other hand, the established sclerotic lesions are resistant to therapies and occasionally leave ugly appearance, therefore early diagnosis and systemic therapy is required. We report a case of morphea with dark pigmentation that mimic malignant melanoma.

CASE REPORT

Twenty-six-year-old man found a brown spot on his back 7 months before consultation. He visited dermatologist because of enlargement of the lesions, and was referred to our clinic suspicious of malignant melanoma. At the first visit, a blackish brown pigment lesion sized 12 x 15 cm located on the middle back. The pigment lesion has irregular border and color. It surrounded with erythema and the center part of the lesion elastic with slightly hard slightly appearance(Figure 1). Dermoscopy revealed typical pigmentation. He had no sclerotic changes or Raynaud phenomenon of extremities.

Skin biopsy specimen from the pigmented area revealed thick and hyalinized collagen bundle proliferation with marked basal pigmentation with some dermalmelano phages(Figure 2). No tumor cell was identified. The specimen from erythematous lesion revealed dense dermal perivascular infiltration with edema.



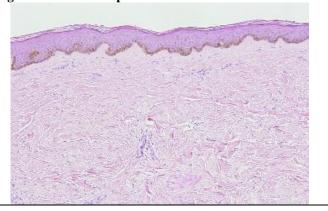
Clinical laboratory examination was within normal limit, and specific auto antibodies for scleroderma including antinuclear antibody, rheumatoid factor, anti-Scl70 antibody, anti-RNA polymerase III antibody and anti-U1-RNP antibody were negative. He was diagnosed with morphea.

The skin lesion was resistant to two weeks topical clobetasol propionate and alprostadil alfadex therapy. Daily oral prednisolone 5mg improved skin lesions, and the sclerotic changes improved leaving pigmentation within two months.

Fig 1. Dark pigmentation with mild sclerotic changes at center and surrounding mild erythema on the back.



Fig 2. A skin biopsy specimen from the glossy dark colored lesion on the back. Thick hyalinized collagen bundles proliferated in the dermis with marked basal pigmentation of the epidermis



DISCUSSION

Morphea (localized scleroderma) is a distinct entity from SSc, and has good prognosis without organ involvement. Morphea develops at any age from children to the elderly [1]. Clinical manifestation of morphea is variable, and divided to 5 types: plaque56%, linear19%, generalized13%, deep11% and bullous. Morphea develops mainly on the trunk and extremities, and face and neck are involved around10% [2].

The diagnosis of morphea in early stage is sometimes difficult. It starts as a red edematous lesion without specific change. Central pigmentation and/or glossy appearance develop lately. Lack of specific laboratory finding sis preventing early diagnosis. Characteristic clinical manifestation of present case is pigmentation. Because of its bizarre form, color variegation and asymmetry, malignant melanoma is suspected and referred to our clinic. Dermoscopy worked very well, and easily ruled out malignant melanoma by the presence of typical pigment network. There were two characteristic findings: presence of lilac ring which is surrounding erythema with violaceous-pink colored erythema surrounding the pigmented lesions, and palpable elastic hardening of the central pigmented lesion. Some

glossy appearance at the centeris also helpful. These are very important findings for early diagnosis of morphea [3].

CONCLUSION

Systemic therapy for morphea is a matter for debate [1]. However, the area of lilac ring responds to low dose predonisolone. In the present case, lilac ring improvedby 5mg of predonisolone leaving mild pigmentation. Already sclerotic lesions are resistant to therapy, however, they became soft with improved pigmentation within two months. Spreading inflammation of morphea to underlying muscle and bone increases the risk of permanent deformity. Because of good response to low dose systemic corticosteroid, early diagnosis and systemic therapy is required for morphea.

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CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest.

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