

Solitary Morphea Profunda with Incidental Acantholysis

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Solitary morphea profunda is a rare form of scleroderma, characterized clinically by a solitary sclerotic plaque, and histologically by marked dermal and subcutaneous fibrosis with an inflammatory infiltrate. We describe another case of this entity presented with an ulcerative, indurated plaque on the left iliac crest, which histologically revealed a focal incidental acantholysis in the overlying epidermis and a marked eosinophilic infiltration through the dermis to the subcutaneous tissue. (*Ann Dermatol* 11(2) 78~81, 1999).

Key Words : Solitary morphea profunda, Incidental acantholysis

Scleroderma occurs in both systemic and localized forms. Localized lesions may be categorized as morphea (localized, guttate, generalized, profunda, and pansclerotic), or linear scleroderma¹. Morphea profunda, a deep type of morphea, appears as a diffuse, deep, bound-down, sclerotic lesion that involves the subcutaneous tissue and fascia². Whitaker et al³ first proposed the term of 'solitary morphea profunda' to describe five patients with a solitary sclerotic plaque mainly on the paraspinal areas, histologically characterized by marked fibrosis, hyalinization of collagen fibers, and inflammatory infiltrates in the deep dermis and subcutis. Thereafter, two additional cases, one under the title of 'solitary fibrosing paraspinal plaque'⁴ and the other associated with perforating osteoma cutis⁵, have been reported. Herein we present a case of a solitary morphea profunda, in which the histological examination showed a focal incidental acantholysis and marked eosinophilic infiltration through the dermis to the subcutaneous tissue.

CASE REPORT

A 24-year-old man presented with a 6-month history of a chronic ulcerative plaque on his left iliac crest. There was no history of trauma or insect bites to the area. He was free of other skin lesions and his previous or family history of skin disorders was unremarkable. Examination revealed a 10 x 6 cm sized, ill-defined, erythematous indurated plaque on the left iliac crest (Fig. 1). The surface was ulcerative and crusted in the center of the lesion and the consistency was hard. Laboratory investigations, including a complete blood count with a differential count, serum eosinophil cationic protein, erythrocyte sedimentation rate, urinalysis, stool examination for parasite eggs, serology tests for syphilis, liver function tests, rheumatoid factor, antistreptococcal antibody titer, and antinuclear antibodies, were all within normal limits or negative. A chest X-ray was normal. Many coagulase positive staphylococci were grown in bacterial cultures from the discharge of the ulcerative lesion. KOH mounting and fungus culture were negative.

Skin biopsies were taken from the edge of the ulcerative lesion. Histological sections showed marked fibrosis through the deep dermis to the subcutaneous fatty tissue, with a polymorphous inflammatory infiltrate of lymphocytes, plasma cells and eosinophils (Fig. 2). One section also showed focal acantholysis, as well as acanthosis, exocytosis of eosinophils and spongiosis in the overlying epi-

Received April 28, 1998.

Accepted for publication August 3, 1998.

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Fig. 1. Ill-defined, erythematous indurated plaque with underlying shallow ulcer and crust on the left iliac crest.

dermis, and edema, vascular proliferation and extravasation of erythrocytes in the papillary dermis and a marked infiltrate of eosinophils in the entire dermis and subcutis (Fig. 3). Periodic-acid Schiff and Ziehl-Neelsen stainings were insignificant. There was no reaction to direct immunofluorescence testing.

Treatment with cefazolin (1 gm/day, i.v.) for 2 weeks gave no improvement and oral triamcinolone (30 mg/day) was added. Three weeks later, when the ulcerative lesion had improved, a re-

Fig. 2. Skin biopsy taken from the edge of the ulcerative lesion shows a marked fibrosis through the deep dermis extending to subcutaneous fatty tissue, with a dense polymorphous infiltrate of lymphocytes, plasma cells and many eosinophils (H & E, $\times 40$).

biopsy was taken from the healing lesion. Compared with previous biopsy findings, the inflammatory

Fig. 3. A: A small acantholytic foci of spongiotic pattern in the overlying epidermis, B: A marked collagen deposition with inflammatory infiltrate, especially of eosinophils in the dermal-subcutaneous junction (H & E, $\times 100$).

infiltrate, especially of eosinophils, was markedly reduced, but marked fibrosis was persistent through the entire dermis to the subcutis. These histological features suggested the possibility of a morphea profunda. A weekly intralesional injection of triamcinolone was started and then the sclerosis in the skin showed some improvement after 2 months of therapy. A follow-up biopsy revealed marked irregular, pigmentary incontinence, chronic inflammatory cell infiltration and marked vascular dilatation, and fibrosis in the dermis and subcutis.

DISCUSSION

Scleroderma can present itself clinically in either a systemic or a localized form. Many variants of scleroderma have been described, but only a few cases of solitary morphea profunda have been reported. The first report of solitary morphea profunda was presented by Whittaker *et al.*¹ in 1989. These authors described five cases with an ill-defined, solitary, indurated plaque which developed on the back, shoulder, neck, or paraspinal area in middle-aged persons. Kirsner *et al.*⁴ preferred the term 'solitary fibrosing paraspinal plaque' to solitary morphea profunda, because the predilection site was paravertebral. Recently, another case associated with osteoma cutis was reported⁵. Histologically, they share common features of marked dermal and subcutaneous fibrosis, with a polymorphous infiltrate including lymphohistiocytes, eosinophils and numerous plasma cells. These histological findings are similar to those of morphea profunda described by Person and Su², but clinically, this entity is distinct from the latter in that it is not generalized or progressive, and not associated with systemic involvement.

Our case showed an ill-defined, erythematous, indurated plaque with chronic superficial ulceration on the iliac crest. One of the previously reported cases also showed an area of ulceration at the center of the plaque as in our case³. The histopathological examination revealed a diffuse dermal and subcutaneous sclerosis, with a polymorphous inflammatory infiltrate of plasma cells, lymphocytes and eosinophils. These findings were consistent with solitary morphea profunda. However, our case is unique in that one section of the biopsy specimen showed a focal acantholysis in

the overlying epidermis.

Focal acantholytic dyskeratosis can be observed occasionally as an incidental histological finding in the epidermis overlying such diverse lesions as dermatofibroma, basal cell epithelioma, melanocytic nevus, and chondrodermatitis nodularis helices, as well as in pityriasis rosea and acral lentiginous malignant melanoma⁶. This finding has been reviewed recently by Sanchez *et al.*⁷ who studied 9,000 skin biopsy specimens. In 14 of these (0.15%), a tiny acantholytic focus was found, either within the lesion or in the adjacent apparently normal skin. The histological diagnoses of these were basal cell carcinoma, keratoacanthoma, psoriasis, elastolytic granuloma, acral arteriovenous angioma, tinea corporis, leukocytoclastic vasculitis. However, to our knowledge, there has been no mention of incidental acantholysis associated with morphea profunda in the literature. The patterns of acantholysis may simulate the pemphigus vulgaris, superficial pemphigus, Hailey-Hailey disease, and unclassifiable acantholysis⁷. Moreover, a fifth "spongiotic" pattern has been described in Grover's disease⁸. According to this classification, our case might be considered as the spongiotic pattern because exocytosis of eosinophils and spongiosis were also seen in the overlying epidermis. Although the exact pathogenic mechanism of incidental acantholysis is unknown, this finding in various skin lesions may most likely represent an unusual reaction of the epidermis to various associated conditions, rather than a specific histological entity. Separation of epidermal cells can be elicited by direct interference with the mechanism of intercellular adhesion ("primary" acantholysis) or by a more general assault on the viability of the cells as a whole ("secondary" acantholysis) as in viral and bacterial skin infections, thermal or chemical trauma⁹. Although we cannot exclude the possibility that a secondary bacterial infection in the ulcerative lesion might contribute to the development of incidental acantholysis in our case, it seems that an eosinophilic inflammatory reaction is partially responsible for this pathological finding because focal acantholysis was incidentally observed only in the specimen with a florid inflammatory infiltrate, especially of marked tissue eosinophilia. Furthermore, the lesion did not respond to antibiotic treatment only, but improved on the introduction of steroid therapy (triamcinolone, oral and intralesional) and follow-up biopsy specimens revealed no

acantholysis with an eosinophilic infiltrate.

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