A Case of Congenital Solitary Morphea Profunda

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A 4-year-old boy has had a solitary sclerotic depressed plaque on the right anterior chest since birth. The histopathologic findings are consistent with morphea profunda: thickening, hyalinization, and homogenization of collagen bundles in the dermis and subcutaneous tissues, admixture with a prominent lymphocytic and plasma cell infiltrate, and sweat glands entrapped between the thickened collagen bundles. We report a case of congenital solitary morphea profunda. (Ann Dermatol 12(4) 306~309, 2000).

Key Words : Solitary morphea profunda, Congenital

Morphea or circumscribed scleroderma is a benign, cutaneous form of scleroderma, that has been clinically classified into guttate, plaque, linear, segmental, and generalized forms[1]. Person and Su[2] described deep morphea, which predominantly involved the subcutaneous tissue in 1979. In 1981, they also reported deep morphea with both subcutaneous and fascial involvement: some of the patients also showed muscular and deep dermal involvement[3]. The deep morphea was characterized by thickening and hyalinization of subcutaneous tissue with a dense inflammatory cell infiltrate[2,3]. The term morphea profunda has been proposed for those with involvement into the subcutaneous tissue and fascia[3].

Whittaker et al.[4] described a series of five adults with an unusual solitary form of morphea profunda as single, fibrotic plaque on the shoulder, back, neck, or paraspinal area that appeared to represent a distinct variant of localized scleroderma. Recently, we reported one case with unusual histologic findings, involving transepidermal elimination of bony material in a patient with solitary morphea profunda, and another rare case with a solitary morphea profunda associated with lymphangiectasia resembling swiss cheese and developed around milia[5,6].

No case of congenital solitary morphea profunda has been previously reported.

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CASE REPORT

A 4-year-old boy was seen at our hospital for evaluation of a sclerotic erythematous plaque on the right chest area. His parents had noted a slightly depressed and mildly hyperpigmented patch at the same site since birth. His lesion was said to have enlarged and thickened with development of erythema. There was no history of trauma or insect bite to the area. The patient complained of no pain or pruritus. He was otherwise in good health and was not taking any medication.

Physical examination revealed an erythematous to brown, nontender, indurated, depressed plaque measuring 4×2.5cm in diameter on the right chest area. The lesion was well defined with an irregular margin. The surface was slightly shiny. Telangiectasia was also noted (Fig. 1). The plaque was not attached to the underlying tissues. His skin showed no other abnormalities, and his general physical

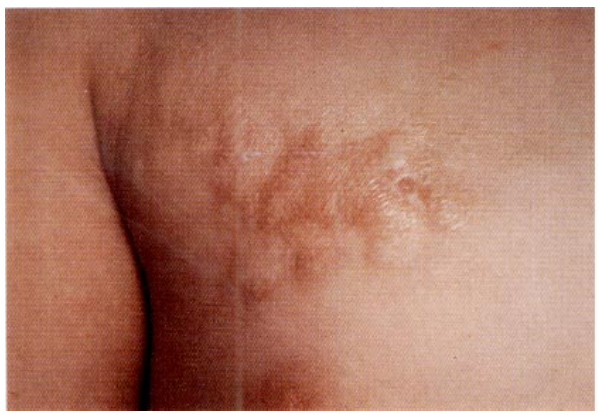

Fig. 1. Right chest of patient with the sclerotic plaque of solitary morphea profunda.

Fig. 2. Extensive thickening, hyalinization, and homogenization of collagen bundles is seen in the deep dermis. Few bound-down eccrine glands are entrapped between the thickened collagen bundles. A moderate inflammatory cell infiltrate within the thickened and hyalinized collagen bundles are seen at the dermis (H & E, ×40).

examination was unremarkable.

A skin biopsy was obtained from the center of the lesion under local anesthesia. A biopsy from the right anterior chest skin showed a normal epidermis with mildly increased melanin pigments in the basal cell layer. The deep dermis and subcutaneous fat were replaced by extensive thickness,

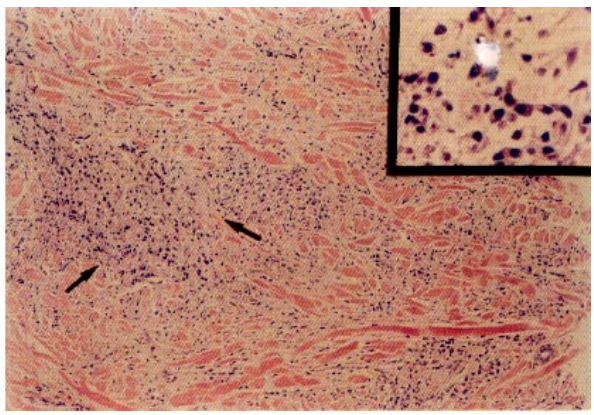

Fig. 3. Inflammatory cell infiltrate consisting of lymphocytes, aggregates of plasma cells in the thickening, hyalinization, and homogenization of collagen bundles (Inset: infiltration of plasma cells). An early lymphoid follicle is formed by inflammatory cell infiltrate (arrows) (H & E, ×100).

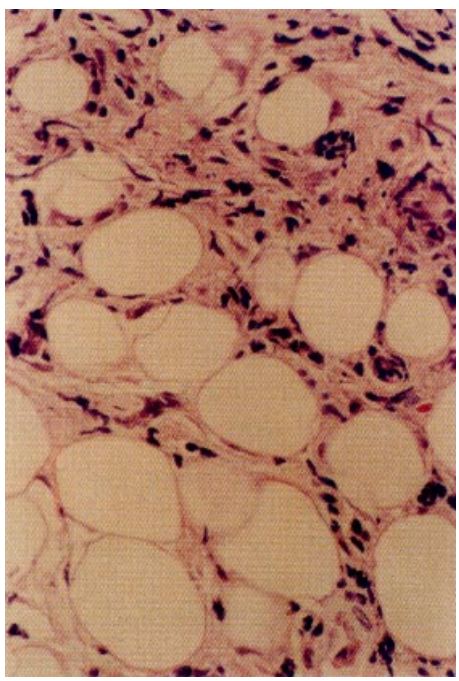

Fig. 4. Inflammatory cell infiltrate is shown in the fibrotic panniculus (H & E, ×400).

hyalinization, and homogenization of collagen bundles. Sweat glands were entrapped between the thickened collagen bundles (Fig. 2). There was a chronic inflammatory cell infiltrate composed of lymphocytes and abundant plasma cells in the dermis consisting of early lymphoid follicles (Fig. 3). This infiltrate extended deeply to involve the dermal-subcutaneous junction, subcutaneous

fat (Fig. 4), and thickened interlobular septae.

There was a slight improvement with topical corticosteroid ointment applied to the lesion over more than three months of observation. The induration of the previous lesion had been slightly decreased, and the depression had been slightly improved.

DISCUSSION

The concept of morphea profunda was first proposed in 1981 by Su and Person[3]. Histologically, the subcutaneous induration consists of extensive thickening and hyalinization of collagen bundles, and inflammatory infiltrates, including lymphocytes, plasma cells, histiocytes, and multinucleated giant cells. The histologic picture resembles those described as localized scleroderma[7]. In contrast to the usual lesions of widespread morphea, the histopathologic changes in morphea profunda include involvement of the deep dermis, subcutaneous fat, and fascia, as well as a more intense inflammatory cell infiltrate. Although plasma cell infiltrates are usually less numerous than lymphocytes in localized scleroderma, a case of linear scleroderma with intense plasma cells infiltrates has recently been reported[8]. Plasma cells are thought to play a role in an immunologic pathogenesis of this condition.

Whittaker et al.[4] described five adults with solitary indurated plaques involving the upper trunk. Although the clinical features in their patients differed from the morphea profunda described by Su and Person[3], the histopathological findings were similar. They concluded that a marked inflammatory response with involvement of the subcutaneous tissue can occur in morphea and that these features are not confined to cases of generalized morphea. The term solitary morphea profunda was proposed for this unusual but distinct clinicopathological entity[4].

Scleroderma may occur in both adults and children, but very rarely in infants[9-11]. And, solitary morphea profunda since birth has not been reported previously.

The clinical differential diagnosis in our patient included keloid, mastocytoma, and a benign or malignant fibrohistocytic tumor. These entities were excluded by the histopathologic findings. The peripheral extensions of the lesion were suggestive of keloid, although the color and depth of the lesion were quite distinct.

Because the skin lesion had existed since birth, it must be differentiated from sclerema neonatorum and subcutaneous fat necrosis of the newborn. Sclerotic skin changes since birth include those due to sclerema neonatorum, subcutaneous fat necrosis of the newborn, and sclerederma. The differentiation of our case from sclerema neonatorum and subcutaneous fat necrosis of the newborn was based upon the presence of thickened homogenized collagen bundles and sclerosis of subcutaneous fat and the absence of needle-like clefts in the subcutaneous adipocytes. In addition, sclerema neonatorum has a fatal course, while our patient demonstrated a benign outcome[12].

The pathogenesis of solitary morphea profunda is not known. Hardening of the skin (sclerodermoid changes) may arise in association with genetic, metabolic, neurogenic, and immunologic disorders, with occupational or chemical exposures, in association with malignancy or as sequelae of infections[1]. However, we are unable to determine the cause of this case. Our patient has no congenital disease, history of medication or exposure to chemical agents.

The prognosis for spontaneous resolution of this disease is poor. Whittaker et al.[4] observed patients from 2 to 17 years. All skin lesions were persistent; they showed no tendency toward progression, and there was no evidence of systemic involvement. Our case had shown no extension or new lesion development since birth. Good therapeutic responses to antimalarial drugs, systemic corticosteroid, and other antiinflammatory agents have been reported[3]. Nevertheless, according to the report of Su and Person[3], none showed complete regression of sclerosis during follow-up of as long as twelve years. In our case, a slight improvement in response to topical corticosteroid ointment in skin lesion was seen after more than three months. The induration of the previous lesion had been slightly decreased, and the depression had been slightly improved.

We experienced a case of solitary morphea profunda developing since birth in a 4-year-boy. To our knowledge, this is the first report of a congenital solitary morphea profunda.

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