Xanthomatous Reaction in Lupus Panniculitis

Sir,

Foam cells are frequently found in Weber-Christian disease or hyperlipemic and normolipemic xanthomas. In 1991, Arai et al. first described the presence of foam cells in alopecia of systemic lupus erythematosus (1) and lesions of discoid lupus erythematosus or systemic sclerosis (2) and termed this change "xanthomatous reaction". Furthermore, they identified the cytoplasmic granules of foam cells as ceroid by histochemical stainings in their cases (3). We here report the first case of lupus panniculitis associated with typical xanthomatous reaction and discuss the possible pathogenetic mechanism.

CASE REPORT

A 17-year-old girl with an ulcerated erythema on her left retroaxillary region was referred to our department in September 1994. She had noticed asymptomatic brownish macules on the flexor aspects of her upper arms in December 1993; however, she had had no systemic manifestations. Physical examination revealed multiple brownish indurated erythemas of walnut-size on the flexor aspects of the upper arms and shoulders. Some of these were concave. In addition, there was an ulcerated erythema with prominent induration on the left retroaxillary region (Fig. 1a).

Peripheral blood and blood chemistry tests were all within normal limits, including myogenic enzymes, amylase and serum lipid levels. Abnormal results were ANA(1:80, homogeneous-speckled type) and anti single-stranded DNA antibody (39.2 units/ml: normal <25). Anti ENA antibodies and anti double-stranded DNA antibody were negative. Histological examination of the indurated erythema of the retroaxillary lesion revealed dense mononuclear infiltrates surrounding the vessels in the dermis and subcutaneous tissue devoid of vasculitis. Focal calcium depositions were also observed in the subcutaneous fat tissue. Severe fibrosis had replaced the subcutaneous fat tissue. Collagen bundles were not hyalinized as seen in morphea. Of particular note was the histological change of the presence of many foam cells among the fibrotic tissue (Fig. 1b). Neither foreign body giant cells nor Touton giant cells were observed. Membranous lipodystrophic change was not seen. Direct immunofluorescence test revealed a linear deposition of IgM limited to the dermoepidermal junction alone. An unstained formalin-fixed section revealed coarse yellow-orange autofluorescent granules in the cytoplasm of foam cells (Fig. 2) when observed under the excitation wavelength of 460–490 nm. This autofluorescence is a characteristic feature of ceroids. On the basis of these clinical and histological findings, we determined the diagnosis of lupus panniculitis with xanthomatous reaction.

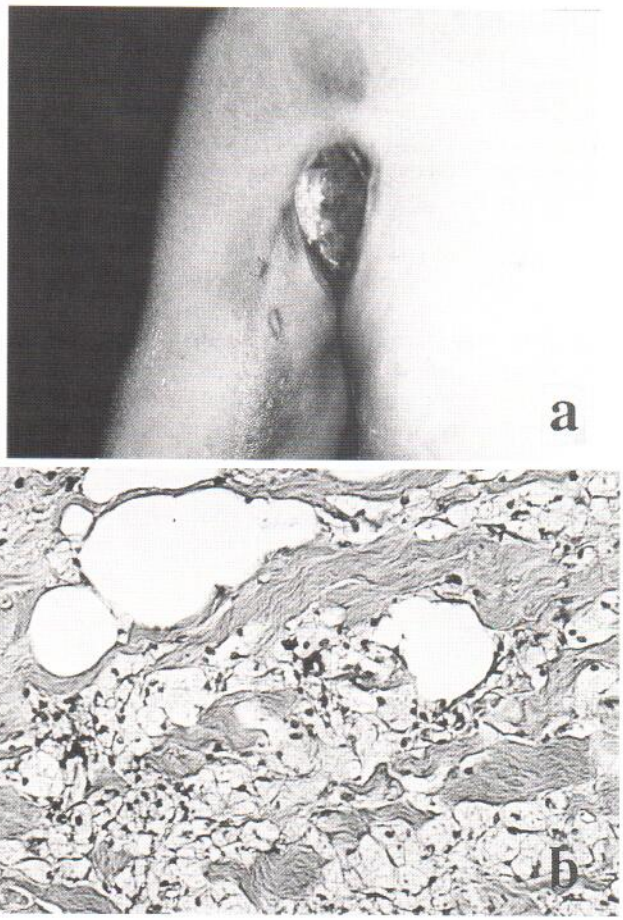

Fig. 1. (*a*) Ulcerated erythema on the retroaxillary area. Indurated erythemas are also present on the shoulder and arm. (*b*) Foam cells in the fibrotic tissue. (Hematoxylin and eosin staining, ×40.)

DISCUSSION

Xanthoma or xanthomatous reaction can occur in association with underlying lymphoproliferative diseases (4). Alternatively, it can result from lipid deposition in damaged or altered skin, such as connective tissue diseases (1, 2), phlebitis/lymphangitis (5), erythroderma from atopic dermatitis (6), etc. In addition, Winkelmann & Oliver proposed a subcutaneous type of adult xanthogranuloma in normal elderly men (7). Thus, xanthomatous reaction may not be a disease-specific change but may represent a certain peculiar reaction

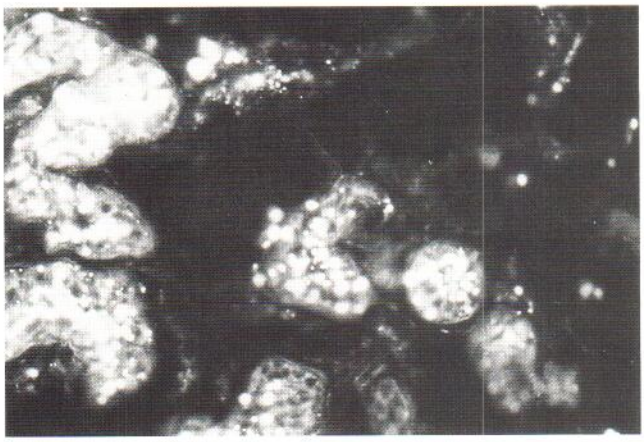

Fig. 2. Autofluorescence in the cytoplasm of foam cells observed under the excitation wavelength of 460–490nm (×400).

to damaged fat tissue. To our knowledge, this is the first case demonstrating xanthomatous reaction associated with lupus panniculitis. We believe that the reaction seen in our patient was a secondary change to lupus panniculitis. Although we retrospectively examined 18 skin specimens diagnosed as lupus panniculitis, only the present case exhibited a xanthomatous reaction. This suggests that this reaction is rare in lupus panniculitis. Either the disease duration or the time of skin biopsy may account for the special nature of this condition.

As a ceroid-related reaction, membranous lipodystrophy is a peculiar type of fat necrosis associated with various diseases such as morphea profunda, lupus panniculitis, ischemic leg ulcer, etc (8). The histological characteristics are multiple microcysts with arabesque-patterned lining and microgranules in histiocytes, and the lining material has proved to be ceroids. Chung et al. (8) suggested the possible role of pressure from fibrotic tissue on fat cells in the development of membranous lipodystrophy. In our opinion, the severe fibrosis and focal calcifications after panniculitis were caused by the disturbance in local circulation. However, the concomitant presence of xanthomatous reaction and membranous lipodystrophy has never been conclusively demonstrated. This may be a result of the lack of attention given to the presence of xanthomatous reaction. We believe, therefore, that both conditions may be generated by a similar physiological reaction, in which unknown factors may contribute to create the histological differences between them.

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