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LETTER TO THE EDITOR

Reticular variant of mid-dermal elastolysis accompanied by persistent urticarial lesions

Dear Editor,

Mid-dermal elastolysis (MDE) is an unusual disease which is histopathologically characterized by selective loss of elastic fibers in the mid-dermis. Clinically, well-circumscribed fine wrinkling and/or discrete perifollicular papular protrusions are usually observed symmetrically arranged on the trunk and proximal aspects of the limbs.[1,2]

A 32-year-old Caucasian woman presented with an over 6-year history of slowly progressing asymptomatic skin lesions. Skin changes appeared first on the upper chest, with insidious extension to the shoulders, and more recently to the epigastric area. Following intense solar exposures during vacations in a tropical country 3 months earlier, she noted rapid spreading of skin changes from the shoulders to the whole back. The clinical picture is demonstrated in Figure 1. Laboratory investigations excluded elevated antinuclear antibodies and antiphospholipid antibodies. Topical treatment with highly potent topical corticosteroids (0.05% clobetasol propionate cream) achieved significant clinical improvement with disappearance of erythema and flattening of papules and plaques especially on breasts and abdomen. However, the fine wrinkles remained unchanged.

Two biopsies were performed, one from urticaria-like lesions under her breasts and one from reticulate erythema on the upper back. Hematoxylin–eosin staining did not demonstrate pathological findings, except for moderate perivascular lymphohistiocytic infiltrates including scattered eosinophils in the superficial dermis and a slight dermal edema in the biopsy specimen taken from her breast lesions. Periodic acid-Schiff and Giemsa stainings were unremarkable. Alcian blue staining excluded increased mucin deposits. In both biopsy sites, van Gieson's staining as well as elastin immunohistochemistry (Fig. 2a) showed selective loss of elastic fibers in the mid-dermis. Negative immunoreactivity was observed for CD117, polymorphonuclear leukocyte elastase and COX-2. Scattered $CD68^+$ cells were observed in the upper dermis in both skin specimens. A moderate to strong immunoreactivity was found for $CD34^+$ cells in the upper and mid-dermis (Fig. 2b), whereas only sparse $CD3^+$, $CD4^+$ and $CD8^+$ lymphocytes contributed to the inflammatory infiltrates.

Urticarial and erythematous papules and/or plaques have only sporadically been reported in MDE patients. In many cases, however, inflammatory skin conditions (e.g. urticaria, atopic dermatitis, granuloma annulare) preceded the occurrence of MDE but did not persist during the course of MDE.[1] Preceding inflammatory skin lesions are also observed in anetoderma, which is a differential diagnosis of MDE frequently associated with antiphospholipid syndrome and systemic lupus erythematosus. However, anetoderma is rather characterized by small lesions whereas MDE occurs in a more diffuse pattern. Though the clinical picture of anetoderma may

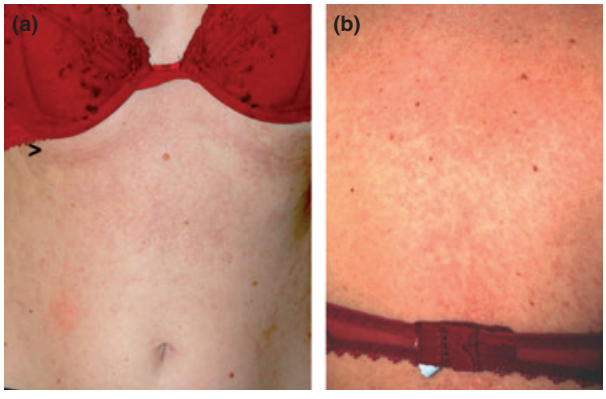


Figure 1. Well-circumscribed areas of finely wrinkled, slightly erythematous skin under her breasts (arrowhead). Furthermore, peau d'orange-like perifollicular protrusions and urticarial, partly annular plaques are seen between her breasts and upper abdomen (a). Moreover, a persistent reticulate erythema with discrete wrinkling is observed on her back (b), shoulders and upper arms.

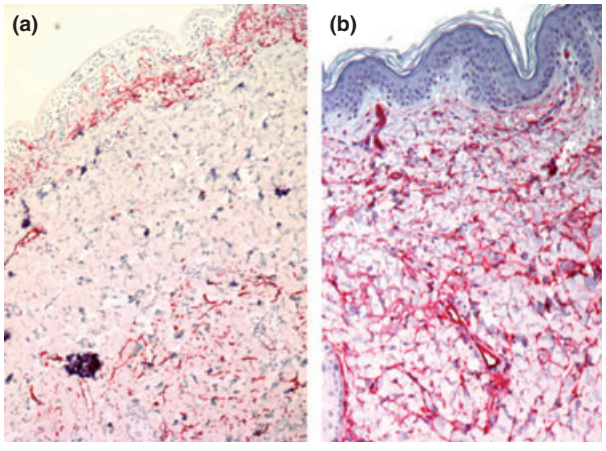


Figure 2. Elastin immunohistochemistry (original magnification ×100) from lesional skin of the abdomen clearly showing selective loss of elastic fibers within the mid-dermis (a). Apart from remarkable $CD34^+$ immunoreactivity in endothelial cells, there was also strong $CD34^+$ immunoreactivity in cells likely corresponding to dendritic fibroblasts and histiocytes in the upper and mid-dermis (b).

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resemble MDE in rare cases, the conditions can be differentiated because histology of anetoderma usually shows loss of elastic fibers in the papillary as well as reticular dermis.[1]

A clinical variant of MDE was first reported in 2001 by Bannister *et al.*[3] who observed fine wrinkles associated with a persistent reticular erythema on the upper trunk of two male patients. Hillen[4] and Martin *et al.*[5] also reported two men who presented with persistent reticular erythema with focal elastolysis in the mid-dermis due to elastophagocytosis by histiocytes and/or macrophages. Although initially regarded as a distinct entity,[3] reticulate erythema with elastophagocytosis very likely falls into the spectrum of MDE.[1,5] Reticular configuration of skin lesions and/or elastophagocytosis by multinucleated giant cells and macrophages have also been observed previously in patients with MDE.[1] The clinical and histological findings of the present case are consistent with the reticular variant of MDE, even though a combination of clinical features and persistence of urticaria-like lesions as shown in the present case has not been documented so far. Data of previous observations indicate that increased matrix metalloproteinase activity, possibly induced by $CD34^+$ cells and followed by elastin degradation, constitute the background on which MDE arises. These mechanisms may be based on predisposing factors and environmental cofactors such as ultraviolet radiation, smoking and sex.[1,2]

In conclusion, the presented case report on a patient with the reticular variant of MDE complicated by persistent urticarial lesions expands the clinical spectrum of MDE and highlights the significance of persistent inflammatory skin alterations during the course of the disease.

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