Mid-Dermal Elastolysis

Brian N. Streams, MD; John M. Williams, MD, PhD; Samuel M. Moschella, MD

Mid-dermal elastolysis (MDE), which presents as fine wrinkling of the skin or perifollicular papules, is extremely rare. This entity is distinguished from other elastolytic disorders by its characteristic bandlike loss of elastic fibers limited to the mid dermis. We report a case of MDE that developed gradually in an otherwise healthy woman without prior cutaneous disease. The current theories on the pathogenesis of MDE also are discussed.

Mid-dermal elastolysis (MDE) is a rare acquired condition that was originally described in 1977 by Shelley and Wood.[1] Localized patches of wrinkled skin following lines of cleavage or perifollicular papules are the 2 usual presentations. Although MDE is believed to be a result of postinflammatory destruction of elastic fibers, many patients have developed MDE without preceding inflammation or other symptoms. We report the case of a patient with a classic presentation of this disorder, supporting the concept that MDE is an idiopathic distinct entity.

Case Report

A 37-year-old white woman, without a significant medical history, presented with a 2-year history of asymptomatic, widespread, well-demarcated but irregularly shaped areas of skin wrinkling. These lesions appeared first on her lower abdomen, slowly spreading to her upper abdomen, chest (Figure 1), flexor surfaces of her upper extremities, upper and middle back, neck, and left distal thigh. Subsequently, she experienced 2 episodes of exercise-induced anaphylaxis. There was no family history of similar lesions, cutaneous disease, or connective tissue disease.

Figure 1. Wrinkling and patulous follicles on the lower chest and abdomen.

Findings from the physical examination revealed soft, skin-colored to lightly erythematous, well-demarcated, atrophic plaques studded with patulous follicles, distributed mainly on the patient's trunk (Figure 2) and lower extremities. A biopsy specimen after Verhoeff-van Gieson staining showed diminished elastic tissue in the middle portion of the dermis, with focal preservation adjacent to dermal appendages (Figure 3). Results of laboratory evaluations (eg, complete blood cell count, serum

Accepted for publication October 9, 2002.
From the Department of Dermatology, Lahey Clinic, Harvard Medical School, Burlington, Massachusetts.
The authors report no conflict of interest.
Presented at the New England Dermatological Society Meeting, Boston, Massachusetts, February 9, 2002.
Reprints: Brian N. Streams, MD, Department of Dermatology, Massachusetts General Hospital, 55 Fruit St, Bartlett Hall 6R, Boston, MA 02114 (e-mail: bstreams@partners.org).

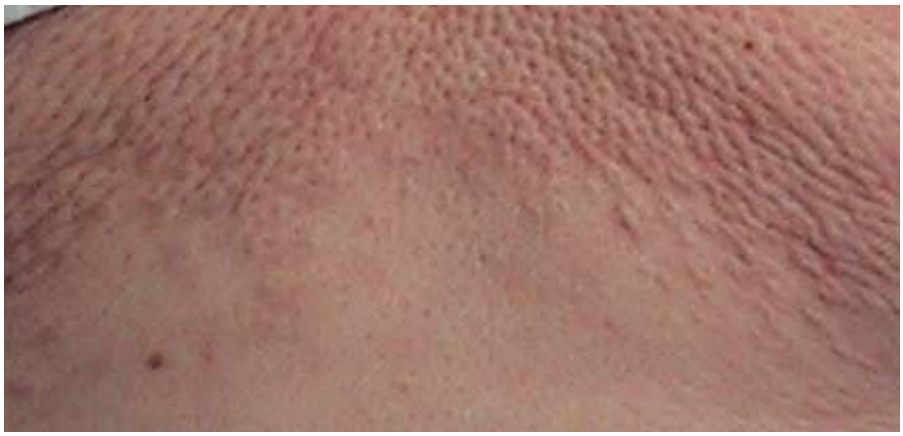

Figure 2. Wrinkling of the skin parallel to skin cleavage lines on the lower chest.

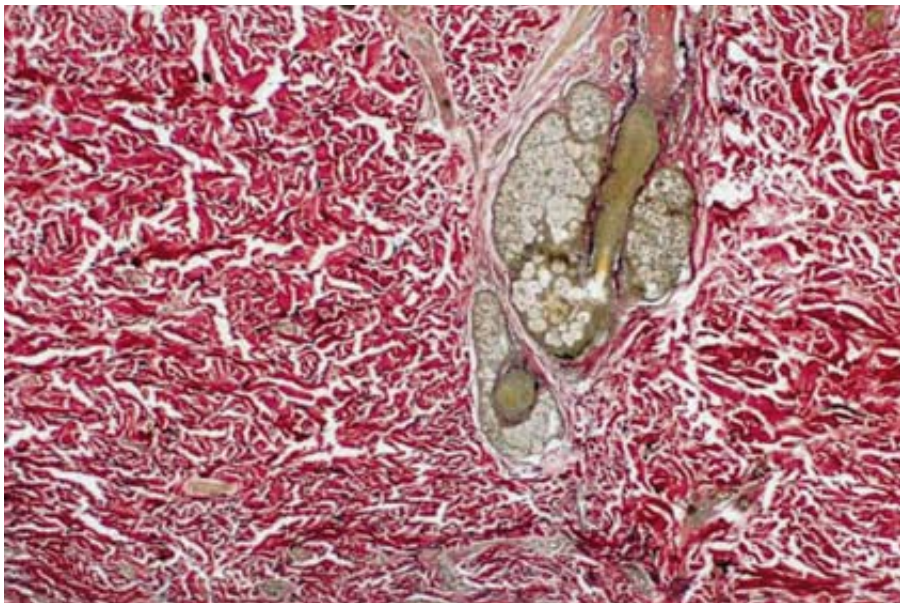

Figure 3. Diminished elastic tissue in the middle portion of the dermis, with focal preservation adjacent to appendages (Verhoeff-van Gieson, original magnification ×10).

electrolytes, erythrocyte sedimentation rate, urinalysis, lipid panel, antinuclear antibody, Sjögren syndrome antibodies) were unremarkable. The patient was treated with colchicine for 2 months without improvement. Her prior medications included an oral contraceptive and sertraline.

Comment

Originally described in 1977 by Shelley and Wood,[1] MDE is a rare, localized, acquired, nonfamilial condition characterized by a bandlike loss of elastic tissue limited to the mid dermis. Although the clinical course is variable, MDE usually affects women in their 3rd to 5th decades of life, appearing as either fine wrinkling of the skin parallel to skin cleavage lines or as perifollicular papules.[2] Lesions are asymptomatic and typically involve the trunk and arms, and rarely, the thighs and face. Most patients report slow progression over several months to years, subsequently reaching a stable stage. Since its initial description, fewer than 20 cases of this entity have been described to our knowledge.[3]

The pathologic features of MDE include the characteristic bandlike loss of elastic fibers limited to the mid dermis. This finding helps differentiate this disorder from other elastolytic disorders, which include cutis laxa (generalized elastolysis), anetoderma (macular atrophy), perifollicular elastolysis, and postinflammatory elastolysis and cutis laxa (intermediate condition consisting of macular atrophy and acquired cutis laxa). In addition, a lymphocytic perivascular infiltrate with histiocytes among collagen bundles may be seen.[4]

Although several theories have been proposed to explain the loss of elastic fibers seen in MDE, the etiology and pathogenesis of this condition remain

unclear. Several lines of evidence suggest a postinflammatory etiology. For example, the first patient described in the literature had a history of urticaria before the onset of wrinkling.[1] Likewise, MDE has been observed in areas of the body previously involved with granuloma annulare and in patients exposed to UV light.[5] Moreover, ultrastructural analyses of MDE cases have demonstrated the presence of phagocytized elastic fibers in mononuclear cells.[4] Similar ultrastructural findings have been reported in acquired cutis laxa, where elastic tissue is absent from the entire reticular dermis and also may be preceded by inflammation or in association with α_1-antitrypsin deficiency. The loss of protection normally provided by this tissue enzyme, which has potent elastase activity, may result in enhanced connective tissue damage by elastases produced by inflammatory cells in the skin. Likewise, postinflammatory elastolysis and cutis laxa, a similar condition reported in Africa and South America, tends to follow an inflammatory phase that may be triggered by insect bites.[6] Many other case reports, however, have described patients without a clear history of prior inflammation, associated dermatologic conditions, or significant UV exposure. Thus, MDE may represent a distinct idiopathic entity or may reflect the end stage or intermediate reaction of multiple inflammatory processes.

Treatment options for MDE are generally disappointing and include colchicine, topical tretinoin (0.01% gel), and topical steroids. Surgical intervention to remove sagging tissue is generally unsuccessful because new folds develop at sites of removal.

REFERENCES

1. Shelley WB, Wood MG. Wrinkles due to idiopathic loss of mid-dermal elastic tissue. *Br J Dermatol.* 1977;97:441-445.
2. Agha A, Hashimoto K, Mahon M. Mid dermal elastolysis: case report and review of the literature. *J Dermatol.* 1996;21:760-766.
3. Sterling J, Coleman N, Pye R. Mid-dermal elastolysis. *Br J Dermatol.* 1994;130:502-506.
4. Neri I, Patrizi A, Fanti PA, et al. Mid-dermal elastolysis: a pathological and ultrastructural study of five cases. *J Cutan Pathol.* 1996;23:165-169.
5. Yen A, Tschen J, Raimer S. Mid-dermal elastolysis in an adolescent subsequent to lesions resembling granuloma annulare. *J Am Acad Dermatol.* 1997;37:870-872.
6. Marshall J, Heyl T, Weber HW. Postinflammatory elastolysis and cutis laxa. a report on a new variety of this phenomenon and a discussion of some syndromes characterized by elastolysis. *S Afr Med J.* 1966;40:1016-1022.