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Case Presentation

Mid-dermal elastolysis: report of a case and literature review

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Abstract

Mid-dermal elastolysis (MDE) is an uncommon and probably underdiagnosed disorder of the elastic tissue. Clinical suspicion and histopathological confirmation are essential for the diagnosis. We report the case of a young woman who presented with this disorder after an inflammatory process during pregnancy and we review the main characteristics of this rare entity.

Keywords:Elastic fibers; Wrinkled patches; Mid dermal

Case synopsis

A 39-year-old woman, without significant past medical history, presented with a several month history of two wrinkled skin plaques on the trunk. The plaques were well circumscribed, flesh-colored and exhibited fine surface wrinkling. The largest was located in the lumbar area (Figure 1) and a smaller similar plaque was on the neck. Physical examination also revealed a plaque on the abdomen that gave the appearance of an "orange peel" (Figure 2). No itch or other symptoms were noted by the patient. The patient noticed the lesions a few months prior to presentation, during late pregnancy and she reported previous erythema after the placement of fetal monitoring.



Figure 1. Wrinkled patch on the lumbar area.



Figure 2. Perifollicular papules on the abdomen.

Two biopsies were obtained from the lumbar and abdominal areas that revealed a normal epidermis with a mild perivascular mononuclear infiltrate in the superficial dermis (Figure 3). Also, there were some interstitial macrophages containing elastic fibers in the mid dermis. Verhoeff-van Gieson stain showed a well-defined area with almost complete absence of elastic fibers in the mid-reticular dermis; the elastic tissue within the papillary and deep reticular dermis was normal (Figure 4). All these findings were consistent with mid-dermal elastolysis.

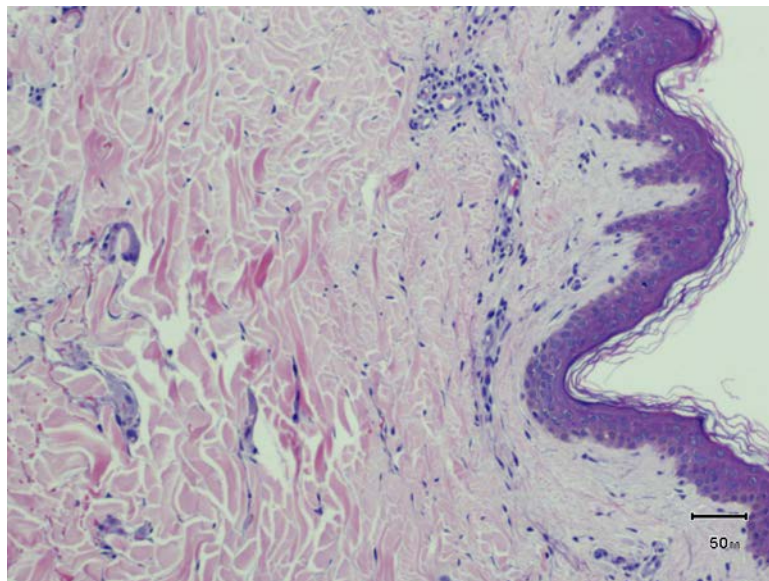
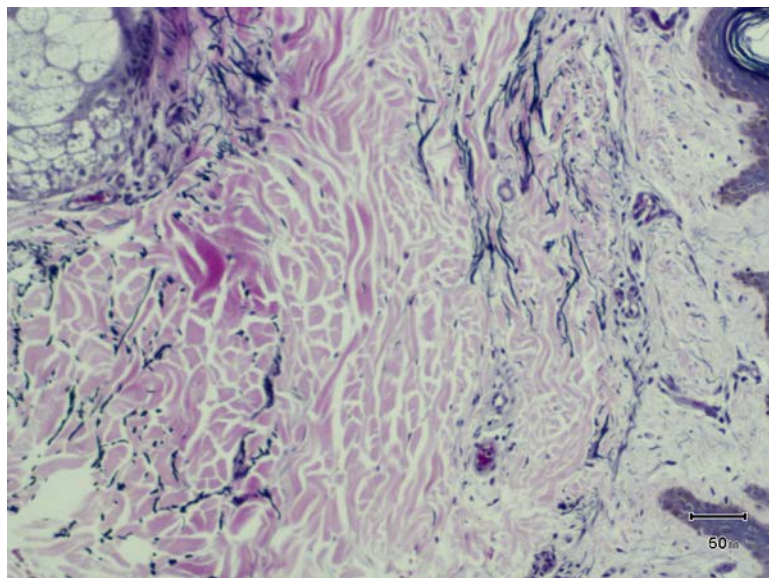


Figure 3. Perivascular mononuclear infiltrate in the superficial dermis and macrophages in the mid dermis (Hematoxylin-eosin, 100X)



Discussion

Mid-dermal elastolysis is an uncommon and probably underdiagnosed acquired skin disorder, characterized histopathologically by decrease or absence of elastic fibers in the mid-dermis. It was first described by Shelley and Wood in 1977 [1]. Since then, about 90 cases have been reported so far, more frequently in middle-aged and Caucasian women.

Clinically, three patterns have been described. Type I is characterized by well-circumscribed, flesh-colored patches with fine wrinkling. Type II consists of perifollicular papular protrusions. These are typically localized on the trunk, neck, and upper limbs, sparing the face, palms and soles, and lower extremities. More recently another clinical variant, type III, consisting of reticular erythema has been described that affects predominantly older men. The lesions are asymptomatic and lack systemic involvement [2].

Pathogenesis remains unknown, although it has been speculated that imbalance in elastin turnover is involved, secondary to environmental factors [3]. Among these, ultraviolet radiation is the major contributing factor. In a high percentage of patients there is a history of excessive sun exposure preceding the development of the lesions. Other authors suggest an autoimmune basis because this entity has been associated with multiple autoimmune disorders, such as systemic lupus erythematosus or Hashimoto's thyroiditis. Moreover, some reported cases were preceded by granuloma annulare [4], urticarial, or acute neutrophilic dermatoses [5]. Thus, it might be the final stage of an inflammatory condition.

Histopathologic confirmation is necessary for the diagnosis of MDE. In early stages, a discrete perivascular lymphocytic infiltrate and elastophagocytosis by macrophages and multinucleated giant cells may be observed. Later, the infiltrate decreases and, with specific staining for elastic fibers, a clear band with absence or decrease of elastic tissue is observed in the middle dermis; the papillary and lower reticular dermis are normal [1-7].

The main entities in the differential diagnosis present with decreased elastic fibers [5-7]. The portion of the dermis involved is the clue to differentiate these. In contrast to MDE, anetoderma and cutis laxa present with a decrease of elastic fibers throughout the entire dermis. Pseudoxanthoma elasticum-like papillary dermal elastolysis only affects the papillary dermis. Furthermore, in perifollicular elastolysis, there is a selective loss of the elastic fibers that surround hair follicles.

No effective treatment has been described for this condition [7]. Multiple therapies have been tried, such as vitamin E, steroids, clofazimine, colchicine, and chloroquine with no significant improvement. Sporadic cases with good response to topical tretinoin and oral dapsone have been published.

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