CLINICAL VIGNETTE

A Case of Mid-Dermal Elastolysis

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Case Report

A 64-year-old Caucasian woman presented with an 8-9 month history of a "rash on the back."

Her past medical history is significant for peptic ulcer disease, osteoporosis, hypertension, and bundle branch block. Medical history is unremarkable for personal or family history of skin cancer. She denies exposure to tanning beds or excessive sun exposure.

Physical examination revealed a large, well-demarcated, faintly erythematous patch with suggestion of crinkling atrophy, encompassing her back from the mid-lumbar region to the sacrum (Figure 1). There were also several scattered, faintly erythematous, somewhat atrophic, approximately ovoid patches on her upper back (Figure 2).

Clinical differential diagnoses included mycosis fungoides and dermatitis. Two biopsies were performed.

Pathologic examination revealed mild superficial perivascular lymphocyte-predominant inflammation with scattered plasma cells and eosinophils (Figure 3). Vierhoff-Van Gieson staining for elastin demonstrated reduced numbers of and fragmentation of elastic fibers (Figure 4). Trichrome stain highlighted normal dermal collagen (Figure 5). Histopathologic diagnosis was mid-dermal elastolysis.

At this point, the patient was referred to Internal Medicine and Rheumatology for further medical work-up. Laboratory testing revealed a negative ANA, SS-A, and SS-B. The patient's IgG, IgA, and IgM levels were all within normal limits.

She was started on triamcinolone 0.1% cream used twice daily and noticed a significant improvement in the appearance of the erythema and pruritus of the lesions within several weeks. Her lesions are now stable, and she has not noticed any new lesions.

Discussion

Mid-dermal elastolysis is a rare, acquired skin condition of the elastic tissue, classically involving the trunk and proximal extremities, and usually sparing the hands and face. Mid-dermal elastolysis is predominantly seen in younger women.[1] Since its first description by Shelly & Wood (1977), there have been around 90 reported cases of mid-dermal elastolysis in the literature.[2] Three chief clinical manifestations have been identified: well-circumscribed fine wrinkles (type I), perifollicular papular protrusions (type II), and rarely, persistent reticular erythema and wrinkling (type III). A definitive diagnosis requires histological band-like loss of elastic fibers along the mid-dermis.[3]

The pathogenesis of mid-dermal elastolysis is unclear. Several studies have found increased elastase synthesis in cultured fibroblasts of patients' affected skin with mid-dermal elastolysis.[1] A few reports described an imbalance between matrix metalloproteinases (MMPs) and endogenous tissue inhibitors of metalloproteinases (TIMPs).[1] Several cases seem to be related to sun exposure and ultraviolet radiation. However, typical histologic actinic changes associated with UV exposure are not seen with mid-dermal elastolysis. In addition, lesions do not typically manifest on chronically UV-exposed sites (e.g., face and back of hands).[1] Interestingly, around ¼ of mid-dermal elastolysis cases were associated with prior use of contraceptives or pregnancy. Smoking was also a shared association between several cases with one report indicating 7 out of 11 patients as smokers.[1]

Several cases have associated mid-dermal elastolysis with autoimmune phenomena (e.g., false-positive serology for *Borrelia burgdoferi*, elevated antinuclear antibodies) and autoimmune disorders (e.g., systemic lupus erythematous, rheumatoid arthritis, Graves' disease, Hashimoto's thyroiditis).[1] It is therefore important to evaluate these patients for possible associated autoimmune disorders.

The diagnosis of mid-dermal elastolysis is compounded by its often non-specific clinical appearance. Clinical differential diagnoses for erythematous, somewhat atrophic patches could include mycosis fungoides, atopic dermatitis, psoriasis, parapsoriasis, early morphea, and pityriasis rosea.

Treatment and management of mid-dermal elastolysis can be challenging. Multiple therapeutic agents have been tried with varying levels of efficacy. Case reports on mid-dermal elastolysis have mentioned the use of agents including topical and systemic steroids, vitamin E, colchicine, clofazimine, chloroquine, topical tretinoin and dapsone.[3]

Soybean extract has also been shown *in vitro* to induce elastin synthesis and inhibit elastase activity, which may have potential therapeutic benefits for elastolytic conditions.[4]

Figures

Figure 1: Large, well-demarcated, faintly erythematous patch.

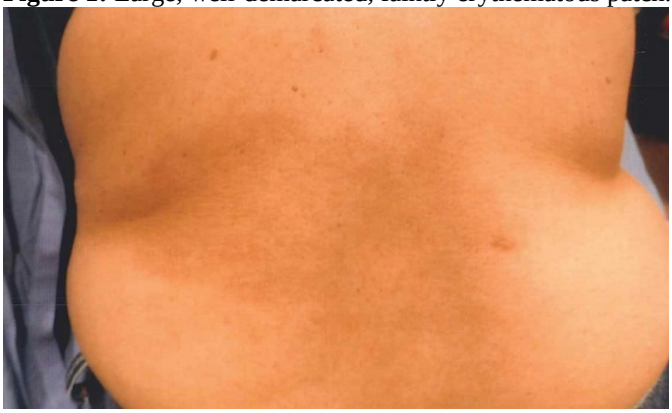


Figure 2: Scattered, faintly erythematous, somewhat atrophic, ovoid patches.

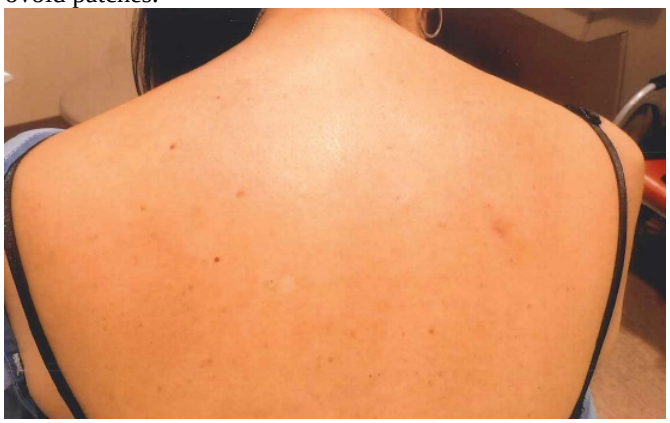


Figure 3: Histologic sections reveal mild, focal lymphocyte-predominant perivascular or interstitial inflammation without an apparent diminution in interstitial fibers *(hematoxylin and eosin, 40x).*

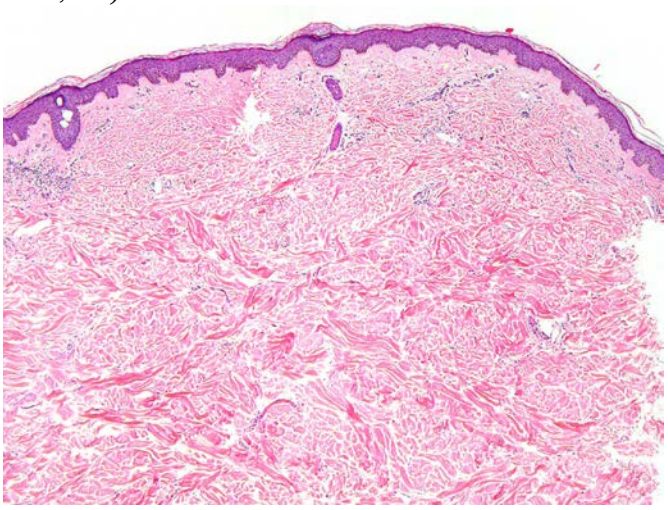


Figure 4: Verhoeff's Van Gieson (EVG) special staining highlights a diminution in mid-dermal black-staining elastic fibers to include shortened and fragmented fibers *(EVG, 100x).*

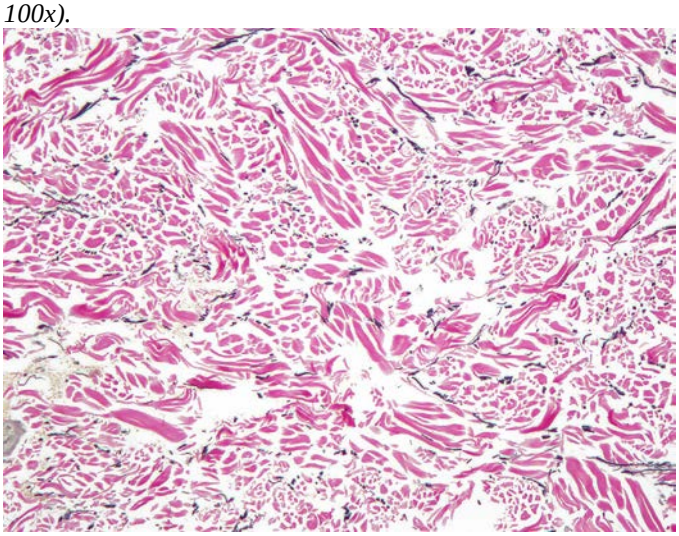
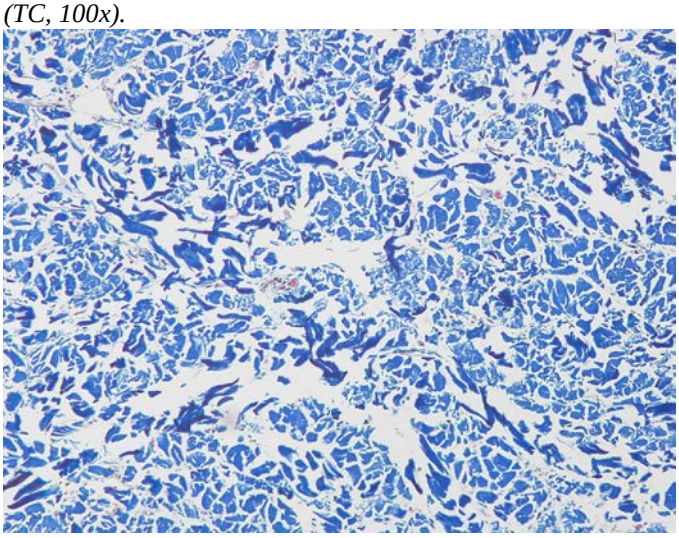


Figure 5: Trichrome special staining highlights an overall normal complement of blue-staining dermal collagen fibers *(TC, 100x).*



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