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Authors
Uldall Pallesen, Kristine Appel
Lindahl, Kim Hein
Bygum, Anette

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Elastosis perforans serpiginosa related to vascular Ehlers-Danlos syndrome

Kristine Appel Uldall Pallesen[1] MD, Kim Hein Lindahl[2] MD, Anette Bygum[1] MD

Affiliations: [1]Department of Dermatology and Allergy Centre, Odense University Hospital, Odense, Denmark, [2]Department of Pathology, Odense University Hospital, Odense, Denmark

Corresponding Author: Kristine Appel Uldall Pallesen MD, Department of Dermatology and Allergy Centre, Odense University Hospital, Odense, Denmark, Tel: 45-50463483, Email: Kristine.Pallesen@rsyd.dk

Abstract

Elastosis perforans serpiginosa (EPS) is a rare skin disease with elimination of connective tissue fibers from dermis to epidermis. The typical presentation shows hyperkeratotic red or skin-colored papules arranged in a circinate pattern. We present a 26-year-old woman with EPS known to have vascular Ehlers-Danlos syndrome.

Keywords: elastosis perforans serpiginos, vascular Ehlers-Danlos syndrome

Introduction

Elastosis perforans serpiginosa (EPS) is a rare perforating skin disorder, which is characterized by papules containing elastic fibers in the epidermis. Elastosis perforans serpiginosa can be idiopathic, related to systemic diseases, or induced by drugs such as D-penicillamine [1]. Vascular Ehlers-Danlos syndrome (EDS) is a rare genetic connective tissue disorder caused by a defect in the production of collagen III, which makes the connective tissue more fragile. We report a case of a patient having both vascular EDS and EPS.

Case Synopsis

A 26–year-old woman with known vascular EDS was referred to our department because of the recent development of skin lesions. Since childhood her skin had been thin and fragile with visible vessels. She bruised easily and wound healing was prolonged with subsequent formation of atrophic scars. Her face was notable for large eyes with narrow lips and nasal bridge. She had an aged look of the hands and feet.

In the last two years before referral, she had developed multiple small hyperkeratotic skin-colored papules arranged in a serpiginous pattern on her hips, thighs, and upper arms (**Figure 1**). The skin

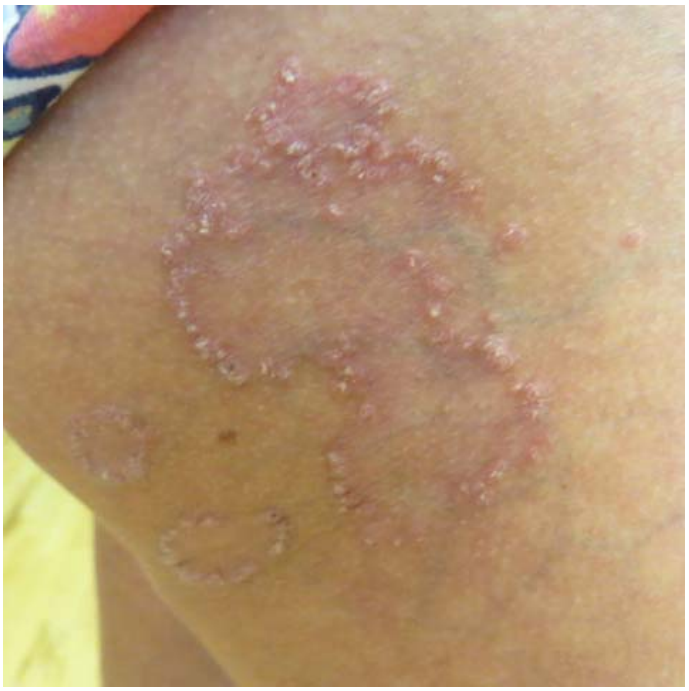

Figure 1. *Elastosis perforans serpiginosa on the leg.*

lesions were treated with potent topical corticosteroids with poor efficacy.

A biopsy specimen of the skin from the hip showed an increased number of coarse elastic fibers in both the papillary and reticular dermis. The epidermis was hyperplastic with hyperkeratosis and several transepithelial, perforating channels filled with eosinophilic elastic fibers and debris, features characteristic of EPS (**Figure 2**).

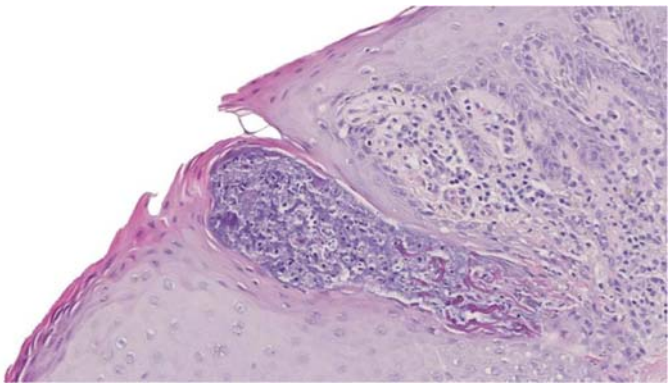

Figure 2. *Histologic image of elastosis perforans serpiginosa. H&E, 100×.*

Case Discussion

Elastosis perforans serpiginosa has been known as a clinical entity since 1953 [2]. The condition is characterized by epidermal elimination of connective tissue fibers through skin papules. The typical presentation of EPS shows hyperkeratotic red or skin-colored papules arranged in circinate patterns. It can be asymptomatic or itchy. Elastosis perforans serpiginosa is more common on the upper extremities, face, and neck but can also develop on the lower extremities. The location on the body does not vary depending upon the particular disease or medication that it has caused. The patients are often young adults and the condition is seen more often in men [3].

Elastosis perforans serpiginosa belongs to the perforating skin disorders also including perforating folliculitis, Kyrle disease, aquired perforating dermatosis, and reactive perforating collagenosis. Three different clinical variants of EPS have been described. The first type is idiopathic. The second type is related to systemic diseases such as cutis laxa, osteogenesis imperfecta, pseudoxanthoma elasticum, EDS, Marfan syndrome, and Down syndrome. The third type is drug induced by medication such as D-penicillamine [3-5].

Ehlers-Danlos syndrome is classified into 6 different types; the vascular type is seen in less than 5%. Vascular EDS is caused by mutations of the *COL3A1* gene. This leads to a reduced amount of mature collagen III [6, 7]. The skin is thin with visible vessels. The vascular type is rare and has a worse prognosis because of the risk of both visceral and arterial rupture [7].

Various treatment modalities for EPS have been reported in the literature, but the treatment response is generally poor. Topical treatment with corticosteroids, tazarotene, tretinoin, calcipotriol, intralesional corticosteroid, narrow ultraviolet B radiation, cryotherapy, and laser therapy have been used.

Conclusion

We present a very rare case of EPS related to the connective tissue disease vascular EDS. Elastosis perforans serpiginosa is a benign disease but can be a challenge to treat.

Potential conflicts of interest

The authors declare no conflicts of interests.

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