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Elastosis perforans serpiginosa: a case successfully treated with intralesional steroids and topical allium cepa-allantoin-pentaglycan gel

Anna Campanati[1], Emanuela Martina[1], Katia Giuliodori[1] ✉, Giulia Ganzetti[1], Barbara Marconi[1], Irene Conte[1], Mirella Giangiacomi[2], Annamaria Offidani[1]

Abstract

Elastosis perforans serpiginosa is a rare skin disease in which abnormal elastic fibers, other connective tissue elements, and cellular debris are expelled from the papillary dermis through the epidermis. Three clinical variants of EPS can be detected: idiopathic, reactive, and drug-induced. Clinically it consists of small horny or umbilicated papules arranged in a linear, arciform, circular, or serpiginous pattern. It usually occurs in young adults and shows a predilection for the head and neck. The lesions are generally asymptomatic or slightly itching. Several treatments have been reported with poor long-term success; these include intralesional and topical corticosteroids, tazarotene, imiquimod, and cryotherapy. We report a case of 40-year-old black woman affected by elastosis perforans serpiginosa that was referred to our department and treated with intralesional injections of triamcinolone acetonide and topical application of allium cepa-allantoin-pentaglycan gel.

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Introduction

Elastosis perforans serpiginosa (EPS) is a rare skin disease generally involving the nape of the neck, face, upper and lower extremities, and trunk, occurring without sex predilection.

Clinically it consists of small horny or umbilicated papules arranged in a linear, arciform, circular, or serpiginous pattern (1). Histologically, it is characterized by abnormal elastic fibers, other connective tissue elements, and cellular debris that are ejected from the papillary dermis through the epidermis.

No treatment of choice can be extrapolated from data in the literature, although several therapies have been proposed to manage patients with EPS.

We describe the case of a 40-year-old black woman affected by EPS that was referred to our department and treated with intralesional injections of triamcinolone acetonide 40 mg/ml and topical application of allium cepa-allantoin-pentaglycan gel.

Case report

A 40-year-old black woman came to our department with a 1-year history of inflammatory lesions on the nape of the neck. Physical examination revealed multiple follicular lesions, confluent into papules and erythematous-crusted plaques, slightly itching, with hypopigmented and atrophic areas, resembling acne keloidalis nuchae (Fig. 1).

A family history was impossible to draw up and the patient's personal history was negative for any significant disease or drug use.

Then we performed a skin biopsy, whose histological examination revealed a thick fibrous dermic band associated with an increase in fragmented elastic fibers and deposits of calcified material, which was ejected over the skin through the infundibular ostium (Figs. 2, 3). The abnormal presence of elastic fibers in the superficial dermis and the appearance of transepidermal elimination suggested a diagnosis of EPS. The presence of calcified material was probably due to a follicular inflammatory process or a traumatic event such as scratching.

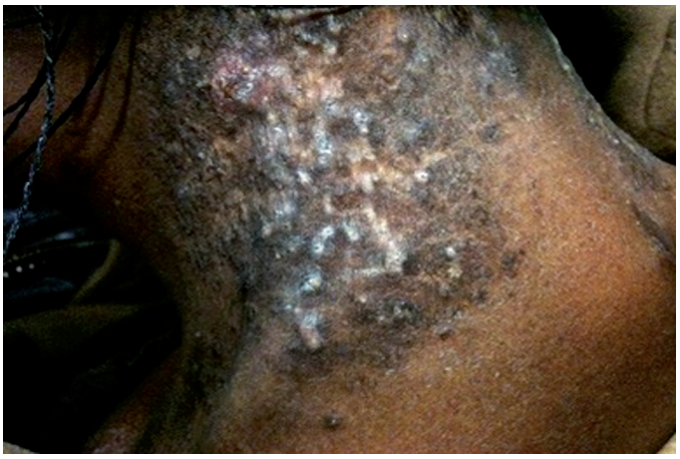

Figure 1 | Patient on presentation to our department with papular eruption on the neck.

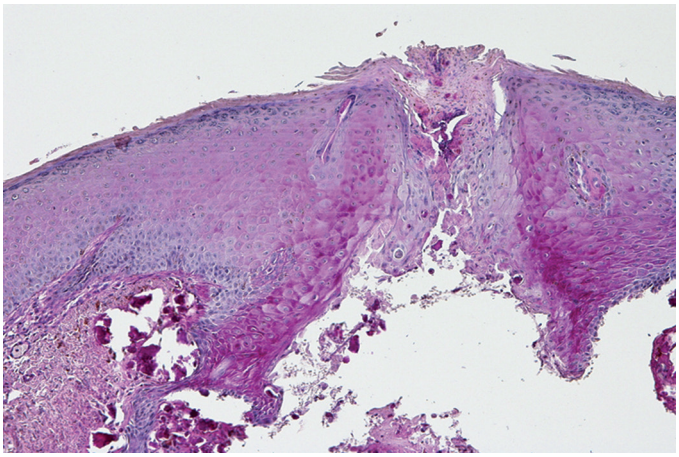

Figure 2 | Biopsy specimen demonstrating transepidermal elimination of altered elastic fibers.

The patient was treated unsuccessfully before the biopsy with oral isotretinoin (0.8 mg/kg/die) for 3 months and, after the histological finding, with high-potency topical corticosteroids (clobet-

[1]Department of Clinical and Molecular Medicine, Dermatology Clinic, Marche Polytechnic University, Ancona, Italy. [2]Department of Neurosciences, Institute of Anatomic Pathology, Marche Polytechnic University, Ancona, Italy. ✉**Corresponding author: katiagiuliodori79@yahoo.it**

asol twice/day for 4 weeks), topical tazarotene (twice/day for 8 weeks), and cryotherapy (three applications for 10 seconds each, every week).

Because multiple therapies had failed, the patient was treated with intralesional injections of triamcinolone acetonide (40 mg/ml) every 15 days for 3 months and with topical application of allium cepa-allantoin-pentaglycan gel twice/day.

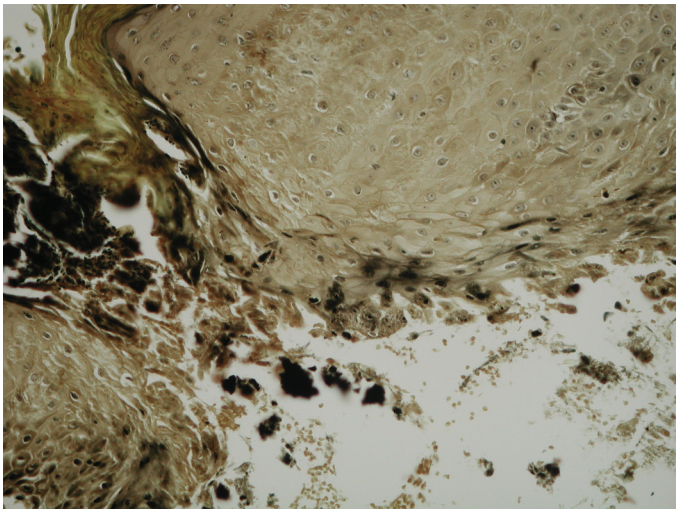

Figure 3 | Biopsy specimen with elastic fiber stain.

After 4 weeks of treatment, the disease was somewhat improved. After 8 weeks of therapy, physical examination revealed a moderate improvement of skin lesions and the patient reported a reduction in itching (Fig. 4).

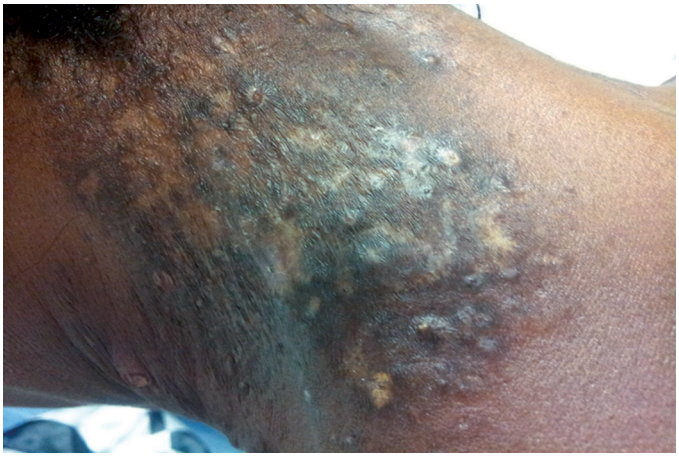

Figure 4 | After 2 months of therapy with intralesional injections of triamcinolone acetonide (40 mg/ml) every 15 days.

Discussion

Elastosis perforans serpiginosa is rare and affects both males and females without racial or geographical preferences; it usually occurs in young adults, even if it can also be observed in childhood or during old age.

In EPS, altered elastic fibers are recognized as non-self material and then extruded through the epidermis (2).

Three clinical variants of EPS can be detected: idiopathic, with a genetic base without any well-known cause; reactive, related to systemic diseases such as Marfan syndrome, Ehlers–Danlos syndrome, Down syndrome, pseudoxanthoma elasticum, and other fibrous tissue diseases (1, 3); and drug-induced, caused by D-penicillamine (4).

Although the pathophysiology of EPS is almost unknown, it has been suggested that a local trigger (biochemical or mechanical) in the dermis could result in the formation of epidermal and follicular channels through which the irritating agent is extruded (5). In most cases, the trigger remains unknown, except for the D-penicillamine–induced forms, because it is a copper chelator able to delay the enzymatic function and the correct deposition of elastic fibers (4).

The molecular mechanism as the basis of the transepidermal elimination of elastic fibers is poorly understood. Optical and electron microscopic analyses showed that the altered elastic fibers generally fill a tortuous channel through the epidermis, and flattened keratinocytes immediately surrounding the perforating channel desquamate directly into the central plug (6).

Fujimoto et al. postulated that the interaction between elastic materials and keratinocytes plays an important part in this extrusion mechanism. They hypothesized that abnormal elastic fibers accumulated in the dermis can be potent inducers of movement and terminal differentiation of keratinocytes via the 67 kDa protein, an elastin receptor. The expression of the 67kDa elastic binding protein has not been reported in normal epidermal keratinocytes, but it can be overexpressed in elastin-rich connective tissues (6).

Several treatments have been described with poor long-term success; they consist of calcipotriene ointment, topical tretinoin, oral isotretinoin (7), glycolic or salicylic acid, topical tazarotene (2), intralesional and topical corticosteroids (8), curettage (1), cryotherapy (9), narrow band ultraviolet B radiation, Er:YAG, CO_2, and dye lasers (10–12).

Differential diagnosis of EPS includes acne keloidalis nuchae, porokeratosis of Mibelli, actinic granuloma, dermatophyte infections, and cutaneous larva migrans.

In our case, we observed papules and nodules of the neck imitating acne keloidalis nuchae and so the patient was initially treated with oral isotretinoin (13-15). After treatment failure, a histological examination was performed to achieve the diagnosis.

Elastosis perforans serpiginosa still represents a clinical and therapeutic challenge, and our experience confirms that intralesional corticosteroids and topical application of allium cepa-allantoin-pentaglycan gel could be an effective therapeutic option (16).

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