Late onset of monomorphous flesh-colored papules of the neck

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Abstract

Papular elastorrhexis is a rare acquired disorder of elastic tissue, characterized by multiple, flesh-colored monomorphous papules usually located on the trunk and the proximal portion of the extremities, occurring in adolescent females. We report a case of PE in an old woman with atypically isolated localization on the neck.

Introduction

Papular elastorrhexis (PE) which was first described by Bordas et al. in 1987, is a rare acquired elastic tissue disorder characterized by multiple asymptomatic, non-follicular, flesh-colored or hypopigmented, monomorphous papules (1, 2). PE is usually located on the trunk and the proximal portion of the extremities in female children or adolescent girls (2). The nosologic position of PE is controversial. Clinical and histological characteristics classify PE as a distinctive entity different from nevus anelasticus and Buschke–Ollendorff syndrome (2,3). The exact etiology of this condition is still unknown. No history of trauma or local inflammation, nor systemic associations or family history are present.

Case report

A 59-year-old woman presented with a one-year history of asymptomatic small papules on the neck. They were slowly increasing in number. The patient had a history of squamous cell carcinoma of the cheek that had been excised. She had not inflammatory dermatosis or trauma on the involved area. There were no similar lesions in her family members. Dermatologic examination showed multiple, 1-4 mm in diameter, flesh-colored, non-follicular, non-confluent and firm papules distributed over the posterolateral sides of the neck (Figure 1).

There were no lesions elsewhere and ophthalmologic examination was normal. Biopsy was performed. Histopathologic analysis showed a normal epidermis and mild thickened collagen bundles in the dermis (Figure 2A). Orcein stains showed a significant loss and fragmentation of elastic fibers in the reticular dermis (Figure 2B). Von Kossa staining was negative. Based on the clinical and histopathological features, the diagnosis of PE was made. We did not prescribe any treatment.

Discussion

PE is a rare disorder of elastic tissue infrequently reported in the literature. The onset is usually within the first or second decade of life with female predominance (75%) (3). The lesions were described as multiple well-defined, soft, hypopigmented or skin-colored, non-follicular oval to round papules with no tendency to group (4). The lesions are symmetrically distributed throughout the chest, abdomen, back and shoulders, upper extremities and rarely thighs (4). There has been no report of extracutaneous manifestation. Uncommon

locations such as involvement of the mandibular, retro-auricular, occipito-cervical regions and face have been reported (5,6). Neck involvement is described in two cases and it was associated to other locations (6,7).

The prominent histopathological feature is reduction, fragmentation or complete loss of elastic bundles in the reticular dermis. A perivascular infiltrate composed of lymphocytes and macrophages in the superficial and deep dermis is present in some cases. Thickened collagen could be seen (8). The pathogenic mechanism of this elastic tissue alteration is unknown, and no local inflammation, trauma or systemic associations have been described. A recent study suggests that abnormal fibroblasts might be involved (9).

Differential diagnosis of PE includes perifollicular elastolysis, middermal elastolysis, pseudoxanthoma elasticum, pseudoxanthoma elasticum-like papillary dermal elastolysis, and white fibrous papulosis of the neck, like in our case.

Oral antibiotics, oral isotretinoin, topical tretinoin, and dibenzoyl peroxide had not been efficient in treating PE. A report has shown anecdotal improvement after intralesional injection of triamcinolone. (5,7,10).

In summary, we report a case of PE in an old woman with atypically isolated localization on the neck.

The diagnosis of PE can be challenging because of the heterogeneous group of elastic tissue disorders. This condition is generally asymptomatic but could cause pruritus and aesthetic discomfort.

Key Clinical Message

* Papular elastorrhexis is a rare acquired disorder of elastic tissue mostly occurring in adolescent females.

* Papular elastorrhexis is characterized by multiple, flesh-colored monomorphous papules usually located on the trunk and the proximal portion of the extremities.

* Neck involvement is described in two cases and it was associated to other locations. We report a case in an old woman with atypically isolated localization on the neck.

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Legend

Figure 1: Multiple flesh-colored non-follicular, 1-4 mm in diameter papules on the posterolateral side of the neck.

Figure 2: (A) Histopathological examination showed hyperplasia of collagen fiber in the dermis (HEX 100). (B) Orcein staining showed a significant reduction and fragmentation of elastic tissue in the reticular dermis (HEX 200).