



Case report

Papular Elastorrhexis: Case report

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Abstract

Papular Elastorrhexis (PE) is a very rare acquired skin disease of unknown etiology characterized by asymptomatic, discrete, hypopigmented, non-follicular, tiny skin papules. We report a 52-year-old Saudi female patient who is otherwise healthy presented with a history of asymptomatic persistent skin lesions on her neck that have been increasing in number since adolescence. There are no similar skin lesions in the family. Skin examination showed multiple tiny non-scaly hypo-pigmented-skin colored dome-shaped papules on her neck. Skin biopsy showed normal epidermis and dermis. The elastic stain showed reduced and fragmented elastic fibers in the reticular dermis.

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Keyword: Papular Elastorrhexis

1. Case report

A 52-year-old Saudi female patient who is otherwise healthy presented with a history of asymptomatic persistent skin lesions on her neck that have been increasing in number since adolescence. There are no similar skin lesions in the family. Past medical, surgical history and systematic review were unremarkable. Skin examination showed multiple tiny non scaly hypopigmented-skin color

dome-shaped papules on her neck (Fig. A.1). Skin biopsy showed normal epidermis and dermis. The elastic stain showed reduced and fragmented elastic fibers in the reticular dermis (Fig. B.1). On the basis of the clinicopathological findings, the diagnosis of Papular Elastorrhexis was made and the patient was reassured.

2. Discussion

Papular Elastorrhexis (PE) is a very rare acquired skin disease. Schirren et al. reported three members of one family with PE, suggesting that it could be a familial disease. However, he proposed that these familial cases could be an abortive form of Buschkee–Ollendorff syndrome (BOS) (Cañueto et al., 2011). BOS is an autosomal dominant disease characterized by disseminated connective tissue nevi and osteopoikilosis. PE was first described by Bordas et al. in 1987 and was considered a variant of nevus anelasticus (Bordas et al., 1987). It commonly occurs dur-

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Figure A.1. Multiple tiny hypopigmented-skin colored papules with smooth surface on the neck of the patient, note some erythematous necrotic lesions due to the effect of electrocautery that has been done as a treatment of these lesions.

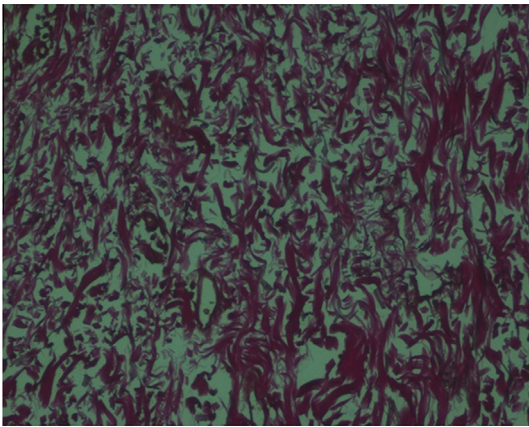


Figure B.1. Elastic stain showing reduced and fragmented elastic fibers in the reticular dermis when compared to a control elastic tissue stain from normal skin of the patient.

ing or after adolescence without any history of trauma or local inflammation (Choi et al., 2011). The exact origin of this condition remains unknown. The disease presents as asymptomatic, discrete, hypopigmented, non-follicular, firm, small papules with a smooth surface. The disease is usually located on the trunk and rarely upper extremities. There has been no report of extracutaneous manifestation (Thomé et al., 2012). Some authors consider PE as a variant of connective tissue nevi (elastoma, collagenoma). However, the histopathological findings are unlike PE. They show increased elastic fibers in elastomas and increased collagen fibers in collagenomas. The

histopathology of PE shows normal skin on hematoxylin and eosin staining, although collagen fibers might be thickened. Elastin stain will show substantial fragmentation or loss of the elastic fibers in the reticular dermis. Electron microscopy may reveal an absolute decrease in elastic fibers, with a relative increase in the fibrillary component of elastic fibers in comparison with normal fibers (Thomé et al., 2012). Clinically, the differential diagnosis of PE includes pseudoxanthoma elasticum, skin tags, and late-onset focal dermal elastosis (LOFDE). However, these conditions have characteristic histopathological findings. Unlike PE, the elastic fibers in LOFDE are increased. In pseudoxanthoma elasticum, there will be a characteristic calcium deposition intermingled with the altered elastic fibers in the dermis.

Some reports have demonstrated that intralesional injections of triamcinolone acetonide resulted in improvement of lesions (Lee et al., 2001; Choi et al., 2004). In one case, papules were flattened by 20 mg/mL of intralesional triamcinolone acetonide (Choi et al., 2004). So far, there is no established treatment for Papular Elastorrhexis.

#### Conflict of interest

The authors have no conflicts of interest that are directly relevant to the content of this case report. No sources of funding were used to assist in preparation of this manuscript.

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