Pseudoxanthoma elasticum-like papillary dermal elastolysis

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KEY WORDS

elastolysis, pseudoxanthoma elasticum-like papillary dermal elastolysis, white fibrous papulosis, white fibrous papulosis of the neck, fibroelastolytic papulosis, mid-dermal elastolysis, pseudoxanthoma elasticum

ABSTRACT

Pseudoxanthoma elasticum-like papillary dermal elastolysis is a rare acquired idiopathic non-inflammatory elastolytic disorder. The skin lesions clinically resemble pseudoxanthoma elasticum but histopathologically there is elastolysis of the papillary dermis and there is no systemic involvement. We report the clinicopathological features of a 67-year-old female with this relatively newly described entity and discuss the pathogenesis, differential diagnosis and controversy regarding nomenclature.

Introduction

Elastolysis refers to loss of elastic fibers. It can affect the papillary dermis, the reticular dermis, or both. Pseudoxanthoma elasticum–like papillary dermal elastolysis (PXE-PDE) is a clinicopathological entity first described by Rongioletti and Rebora in 1992 (1). It is an acquired elastolytic disorder that mainly affects the papillary dermis. Clinically it resembles pseudoxanthoma elasticum (PXE), but it differs histologically and there is no systemic involvement, in contrast to PXE.

Case Report

A 67-year-old Kuwaiti woman presented with asymptomatic skin lesions of 4 years' duration affecting the sides of the neck. She denied any preceding trauma, inflammation, urticaria, or history of sun exposure at the affected sites. There was no history of medical problems and she was on no medications. Examination of the skin revealed multiple 3 to 4 mm skin-colored to yellowish, soft, closely set, cobblestone-patterned papules. The lesions were symmetrically distributed on the sides of the neck (Figure 1). Results of the complete blood cell count, biochemical profile, urinalysis, and stool examination were within normal limits. Chest X-ray and cardiovascular investigations were non-contributory and ophthalmological examination did not reveal angioid streaks or other retinal changes. Histological examination with hematoxylin and eosin revealed only flattened epidermis and a sparse perivascular dermal infiltrate (Fig. 2). Using a special stain for elastic fibers (Verhoeff-van Gieson), a marked band-like reduction of elastic fibers in the papillary dermis that was focally absent was observed (Fig. 3). Mild reduction of elastic tissue in the reticular dermis was also noted. No calcification was seen using von Kossa staining.

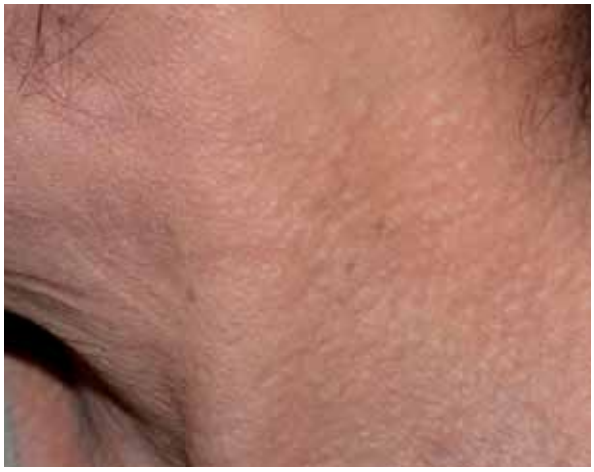

Figure 1. Multiple papules on the side of the neck.

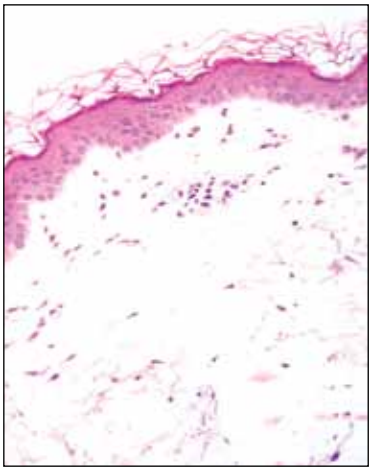

Figure 2. Almost normal skin (Hematoxylin-eosin stain; original magnification 400×).

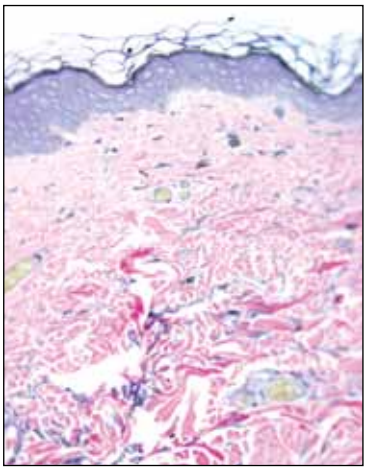

Figure 3. Marked decrease and fragmentation of elastic fibers in the papillary dermis (Verhoeff-van Gieson stain; original magnification 400×).

Discussion

Since the first description by Rongioletti and Rebora in 1992, around 20 cases of PXE-PDE have been reported in the medical literature (1–13). PXE-PDE is a rare, acquired condition characterized by non-follicular yellowish papules coalescing into plaques resembling PXE; it is generally localized on the sides of the neck and subclavicular regions in elderly persons and by loss of or decrease in elastic fibers in the papillary dermis (8).

All the cases reported to date have occurred in women and almost all of them occurred in elderly people around 60 or older (1–13). Thus, any hypothesis regarding the etiology of the disease should explain why it mainly occurs in elderly women.

Although the pathogenesis of PXE-PDE remains poorly understood, ultraviolet (UV) radiation, intrinsic aging, or abnormal elastogenesis may be implicated (2, 4, 14, 15).

Ultraviolet radiation may be involved, but it cannot explain the lesions in our patient because, although the neck is usually sun exposed, our patient had worn the hijab since adolescence. Orlandi et al. made the same observation in two nuns (6). Moreover, sometimes sun-protected skin is also affected (2, 4).

Intrinsic aging seems to be an attractive explanation, especially considering that PXE-PDE mostly occurs in women after their fifth decade. It has been suggested that, because elastin and fibrillin-1 were lost in PXE-PDE patients, it is likely that intrinsic aging is one of several mechanisms involved in its pathogenesis (12). Here, however, another issue is raised; intrinsic aging occurs in both men and women, so why are all cases of PXE-PDE reported in women? It is postulated that women are more concerned about their appearance than men and thus seek medical advice for cosmetic reasons more often (7).

Moreover, there have been two reports from Korea of the condition occurring in 41- and 26-year-old women (10, 16). Therefore intrinsic aging, although important, may be not the only cause.

Pseudoxanthoma elasticum (PXE), mid-dermal elastolysis (MDE), and white fibrous papulosis of the neck (WFPN) should be considered in the differential diagnosis of PXE-PDE.

The average age of onset of PXE is 13 years and it has systemic involvement, characterized by fragmentation and calcification of the elastic fibers of the skin, the retina (angioid streaks), and the cardiovascular and gastrointestinal systems (13).

In MDE, clinically there is diffuse wrinkling, small white papules or perifollicular papular protrusions with a "peau d'orange" appearance, or persistent reticular erythema and histological absence of mid-dermal elastic fibers and sometimes phagocytosis of elastic fibers (17).

WFPN is characterized by multiple confluent white or yellowish papules mainly on the neck (18). Clinically they resemble PXE-PDE. The main pathological feature of WFPN is thickened papillary dermal collagen with decreased elastic fibers (18).

Some authors suggest the term fibroelastolytic papulosis (FEP) of the neck to encompass both PXE-PDE and WFPN (14, 15).

In conclusion, we think that PXE-PDE, WFPN, and fibroelastolytic papulosis are all the same and, until more research indicates otherwise, they probably represent an aging response.

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