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# Pseudoxanthoma Elasticum-like Papillary Dermal Elastolysis

## *A Report of Two Cases and Review of the Literature*

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**Pseudoxanthoma elasticum-like papillary dermal elastolysis is a peculiar idiopathic elastolytic disorder with cutaneous lesions clinically resembling pseudoxanthoma elasticum with partial or total band-like elastolysis of the papillary dermis histopathologically, and without systemic complications. We here report 2 new cases and review the clinicopathological features of patients with this diagnosis in the literature. The possible pathogenesis of this recently described entity is discussed.** ***Key words: papules; neck; age; elastic fibres.***

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In 1992 Rongioletti & Rebora (1) introduced the term pseudoxanthoma elasticum-like papillary dermal elastolysis, describing a peculiar dermatosis which combined lesions resembling pseudoxanthoma elasticum (PXE) clinically, and elastolysis confined to the papillary dermis histopathologically, without systemic involvement. We here report 2 cases of PXE-like papillary dermal elastolysis with the typical clinical features of this new entity.

## CASE REPORTS

### *Case 1*

A 68-year-old woman had a 4-year-history of multiple yellowish non-follicular papules, 2 to 3 mm in diameter, with a symmetrical distribution affecting the supraclavicular regions, the sides and the posterior region of the neck (Fig. 1). She had an essential hypertension, for which she was being treated with a thiazide diuretic (Indapamide), and hypercholesterolaemia. Histological examination with hematoxylin-eosin stain revealed only a flattened epidermis. On special stain for elastic fibres (Verhoff-van Gieson) there was a band-like loss of elastic fibres in the papillary dermis. von Kossa stain did not disclose any calcifications. Complementary studies were within normal limits, except for a mild hypercholesterolaemia. Ophthalmological examination revealed no angioid streaks and there was only an incomplete right branch blockade on electrocardiogram. Chest X-ray and vascular examination were not contributory.

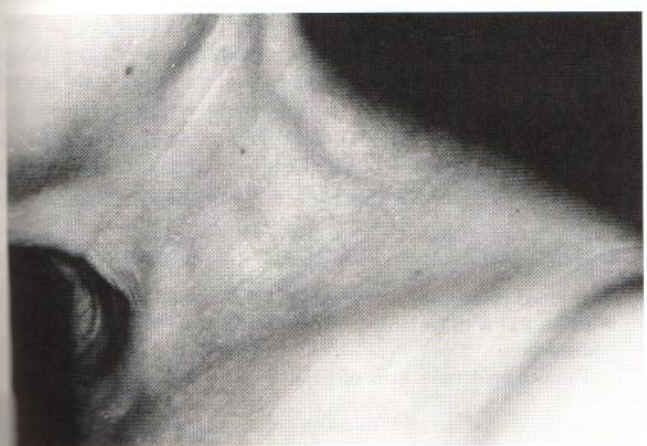

*Fig. 1.* Patient 1. Multiple yellowish papules arranged in a cobblestone-patterned plaque on the supraclavicular region and neck.

### *Case 2*

A 70-year-old woman with a long-standing history of essential hypertension and hypercholesterolaemia complained of mild pruritic but otherwise asymptomatic lesions of unknown evolution. On physical examination there were multiple skin-coloured to yellowish cobblestone-patterned papules, which were symmetrically distributed on the supraclavicular regions and on the sides of the neck. A skin biopsy with van Gieson stain showed a marked band-like reduction of elastic fibres confined to the papillary dermis, with the elastic network in the reticular dermis being unaffected (Fig. 2). No dermal calcifications were observed with the von Kossa stain. Laboratory

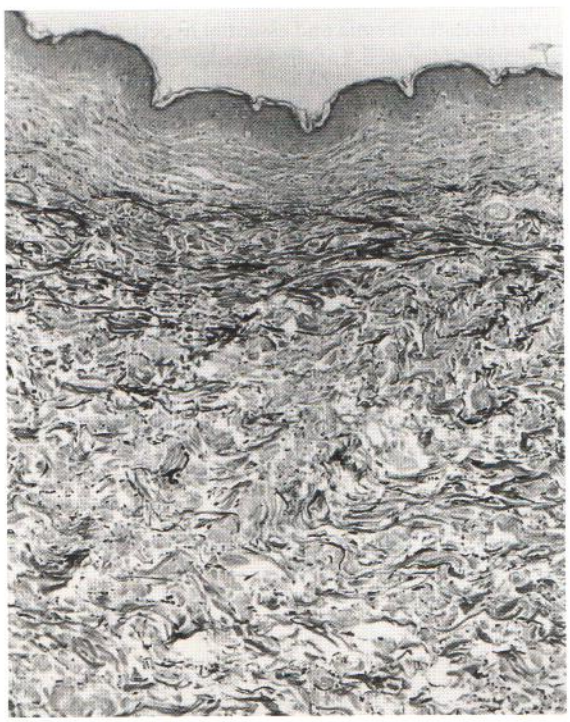

*Fig. 2.* Patient 2. Loss of elastic fibres in the papillary dermis. The elastic network in the reticular dermis is preserved (Verhoff-van Gieson stain × 40).

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studies disclosed only a mild hypercholesterolaemia. No angioid streaks were seen on ophthalmological examination. Chest X-ray, electrocardiogram and vascular examination were within normal limits.

## DISCUSSION

Our 2 patients developed skin lesions typically described for PXE, with elastolysis confined to the papillary dermis and without systemic involvement, a new condition first reported in 1992 by Rongioletti & Rebora, who proposed the descriptive term of PXE-like papillary dermal elastolysis. Since then at least 6 cases have been reported in the international literature (Table I). PXE-like papillary dermal elastolysis affects mainly women in late adulthood (63–80 years) in the form of yellowish non-follicular papules symmetrically distributed on the sides of the neck, flexor aspect of forearms and the lower abdomen. Skin lesions are asymptomatic or mildly pruritic (2, present case 2) and appear progressively in a period of time ranging from 4 months to 5 years. No history of inflammation, trauma or prolonged sun exposure precedes the development of cutaneous lesions. Histopathologically, the hallmark of PXE-like papillary dermal elastolysis is a partial or total band-like loss of elastic fibres in the papillary dermis, although elastolysis may affect the reticular dermis with involvement of the perifollicular region (1). In addition to the reduction of elastic fibres, they may also be seen clumped and fragmented (2). In 2 patients, ultrastructural examination of the involved skin revealed the presence of immature elastic fibres and some fibroblast-like cells with dilated cisternae in their rough endoplasmic reticulum (1, 3). No signs of solar elastosis were observed, and only one case showed a mild lymphocytic infiltration distributed around superficial capillaries (4). Our 2 patients have some clinical features in common. Both were unrelated nuns in late adulthood, with a long-standing history of hypercholesterolaemia and essential hypertension. Although hypertension is a well-known vascular complication in PXE due to renal artery involvement, it has only been described in one patient with PXE-like papillary dermal elastolysis (2), and as for hypercholesterolaemia, its occurrence may be merely coincidental, since there is a high prevalence of these diseases in an aged population where PXE-like papillary dermal elastolysis develops. No history of prolonged sun exposure, especially on the neck, was obtained from our 2 patients, as could be assumed from the lack of indirect signs of actinic elastosis in biopsies from lesional skin, making improbable a specific role of actinic radiation in the aetiology of PXE-like papillary dermal elastolysis. The main differential diagnosis of PXE-like papillary dermal elastolysis has to be made with pseudoxanthoma elasticum, in which one observes fragmentation and calcification of elastic fibres affecting mainly the reticular dermis, and systemic involvement in the form of angioid streaks or splits in Bruchs's membrane and vascular calcifications leading to either ischaemic or haemorrhagic potentially serious complications (5). Many other acquired elastolytic disorders may simulate PXE-like skin lesions (Table II) but unlike PXE-like papillary dermal elastolysis, they lack the superficial band-like elastolytic pattern as single histological feature. Two clinicopathological entities deserve special mention due to the clinical or histological resemblance to PXE-like papillary dermal elastolysis: the recently described white fibrous papulosis of the neck (WFP) (12–15) and the non-inflammatory type of mid-dermal elastolysis (16). WFP was first described in 1985 in 32 Japanese patients, although it has also been reported in Europeans. The clinical picture of WFP may evoke PXE-like lesions with multiple, non-follicular asymptomatic papules distributed mainly around the neck in elderly persons (most of the patients are males). Nevertheless, skin lesions of WFP are not confluent, lacking a cobblestone pattern, are rather pale than yellowish in colour, and may affect the trunk. Although elastolysis of the papillary and reticular dermis has been reported (13, 14), the major histological finding of WFP is a thickening of collagen bundles in

Table I. *Clinical features of patients with PXE-like papillary dermal elastolysis*

EF: elastic fibres, PD: papillary dermis, RD: reticular dermis, ND: not done, UI: ultrastructural investigation.

| Patient | Age/Sex | Lesions | Evolution | Histology | UI |
|---|---|---|---|---|---|
| Rongioletti & Rebora, 1992(1). | 65 years Female | Supraclavicular, mastoidal, laterocervical. | 5 years | Loss of EF in PD. Slightly reduced in RD. | Immature EF in upper RD. |
| | 63 years Female | Supraclavicular and cervical. | 4 years | Loss of EF in PD. Slightly reduced in RD. | ND |
| El-Charif et al., 1994(2). | 80 years Female | Cervical, forearm, lower abdomen. | 4 months | Atrophic epidermis. Reduction and fragmentation of EF in PD. | ND |
| | 63 years Female | Sides of the neck. | 1 year | Loss of EF in PD. EF clumped and fragmented at the junction of PD and RD. | ND |
| Patritzi et al., 1994(3). | 69 years Female | Sides of the neck. | 4 years | Loss of EF in PD. | Immature EF in RD. |
| Pirard et al., 1994(4). | 73 years Female | Sides of the neck, supraclavicular. | 1 year | Thinned epidermis. Loss of EF in PD. EF in RD with variable thickness and distortion. | ND |
| Present case 1. | 68 years Female | Posterior and lateral neck. Supraclavicular. | 4 years | Thinned epidermis. Loss of EF in PD. | ND |
| Present case 2. | 70 years Female | Laterocervical and supraclavicular. | Unknown | Loss of EF in PD. | ND |

Table II. *Acquired elastolytic disorders*

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| Postinflammatory elastolysis and cutis laxa (6) |
| Anetoderma (7) |
| Cutis laxa (8) |
| Perifollicular elastolysis (9) |
| Amyloid elastosis (10) |
| Generalized elastolysis (11) |
| White fibrous papulosis of the neck (12–15) |
| Mid-dermal elastolysis (16, 17) |

the papillary to mid-dermis. Mid-dermal elastolysis (MDE) was first described as "wrinkles due to idiopathic loss of mid dermal elastic tissue" (17), the aetiology of which was related to a preceding inflammatory process. Brenner et al. (16) described an additional morphological pattern of MDE (type II, non-inflammatory MDE) consisting of tiny perifollicular papules, which could resemble PXE-like skin lesions clinically but which differed in the perifollicular distribution and a diffuse loss of elastic fibres throughout the mid-dermis.

As far as the aetiology of PXE-like papillary dermal elastolysis is concerned, different factors have been implied (UV radiation, abnormal elastogenesis and aging). Both conditions, WFP and type II MDE, have been related, along with PXE-like papillary dermal elastolysis, to a common aetiopathological factor of intrinsic or chronological aging (in contrast to extrinsic or actinic-related aging), due to the similar histological and ultrastructural findings that they share with temporary wrinkles, a physiological phenomenon of aging observed on sun-protected skin (18). Hashimoto & Tye (19) reported the case of an 86-year-old woman who developed a clinical picture of pruritus followed by the development of multiple yellowish non-confluent papules on the neck, shoulders, upper chest and back. Histological findings were similar to those described for PXE-like papillary dermal elastolysis, and electron microscopic investigation demonstrated severe elastic fibre abnormalities with disorganization of elastofibrils, diminished deposition of elastin and engulfment of abnormal elastic fibres by dermal phagocytes. Although they referred to this case as "upper dermal elastolysis" and believe that it differs from PXE-like papillary dermal elastolysis because of the lack of arrangement of papules in plaques, a less yellowish colour and involvement of the chest and back (cutaneous manifestations which resemble those of WFP), it is very likely that this patient could also be fitted into the same clinicopathological picture of dermal elastolysis. Thus, PXE-like papillary dermal elastolysis, white fibrous papulosis of the neck, temporary wrinkles and the non-inflammatory type of mid-dermal elastolysis could be lumped together in a "non-inflammatory dermal elastolytic syndrome", with a spectrum of clinical presentations depending on the location and extension of elastic fibre reduction, as well as on involvement of other connective tissue components (i.e. collagen bundles). Whether those diseases represent merely a consequence of an age-related process of elastic fibre degeneration (in which case a greater incidence in elderly population would be expected), or their occurrence lies in a primary defect in the elastic fibre components, requires further investigation.

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