Fibroelastolytic Patterns of Intrinsic Skin Aging:
Pseudoxanthoma Elasticum-like Papillary Dermal Elastolysis and White Fibrous Papulosis of the Neck

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Pseudoxanthoma elasticum-like papillary dermal elastolysis is characterized by multiple whitish-yellow papules on the neck and supraclavicular regions of elderly persons. The papules resemble those in pseudoxanthoma elasticum. There is also an histological loss or decrease of the number in elastic fibers in the papillary dermis. There is no family history of such skin lesions and lack of cardiovascular or ocular abnormalities. White fibrous papulosis of the neck is characterized by several whitish to yellowish, tiny, firm papules located on the posterior or lateral sides of the neck in the elderly. We herein report a case of pseudoxanthoma elasticum-like papillary dermal elastolysis and another typical case of white fibrous papulosis of the neck. According to the literature, both diseases are best viewed as two fibroelastolytic patterns of intrinsic skin aging. (Dermatol Sinica 21: 402-407, 2003)

Key words: Pseudoxanthoma elasticum-like papillary dermal elastolysis, White fibrous papulosis of the neck, Intrinsic skin aging, Fibroelastolytic papulosis of the neck

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INTRODUCTION

Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) and white fibrous papulosis of the neck (WFPN) share several similarities. These include location, mainly the sides of the neck, multiple papular eruptions, histological changes in skin aging, and the loss of elastic fibers in the papillary dermis. They are considered to be two fibroelastolytic patterns of intrinsic skin aging.

In this report, we present two typical cases of PXE-PDE and WFPN and compare the clinical and pathological features of both diseases.

CASE REPORT

Case 1

A 57-year-old woman had a 3-year history of slowly progressive, symmetric, asymptomatic skin lesions on the sides of her neck, the axillae and the upper back. She denied any previous urticarial or inflammatory disorders. There was also no history of preceding trauma or prolonged sun exposure at the affected sites. Dermatological examinations revealed many white-yellow, non-follicular papules coalescing into plaque with a cobblestone-like pattern on her laterocervical, mastoidal and supraclavicular regions (Fig. 1). None of her family members had any similar skin manifestations. The patient's medical history was unremarkable, and the skin lesions were not related to the medications she had taken. Laboratory tests, chest X-ray study and cardiovascular investigations were not contributory and ophthalmological examinations did not reveal angioid streaks or other retinal changes.

A biopsy from the involved skin did not reveal any abnormalities under haematoxylin

Fig. 1

There are multiple yellowish, non-follicular, soft papules coalescing into plaques with a cobblestone-like appearance on both sides of the neck and supraclavicular regions. (A) the anterior view; (B) the posterior view; (C) the close view of the lesions.

Fig. 2

Complete loss of the elastic fibers in the papillary dermis (orcein stain).
and eosin (H & E) stain, but a complete band-like loss of elastic fibers in the papillary dermis along with focal elastogenic change in the sub-papillary or mid-dermis under the orcein stain were noted (Fig. 2). She was diagnosed as having PXE-PDE.

**Case 2**

A 42-year-old woman had a 4-year history of asymptomatic skin lesions on her neck. She denied any trauma history or other preceding cutaneous disorders. Dermatological examinations revealed six, discrete, ricegrain-sized, yellowish to whitish, round or oval papules on the left side of her neck (Fig. 3). Two similar tiny whitish to yellowish papules were also noted on the right side of the neck. They were unrelated to any hair follicles and were not pedunculated (Fig. 3). Histological examinations with a H & E stain did not show any remarkable epidermal change with a relatively circumscribed area consisting of thickened collagen bundles in the papillary to mid-dermis (Fig. 4). Above this zone of abnormal collagen, there was a decrease in the elastic tissue in the upper dermis under the orcein stain (Fig. 5). The diagnosis was WFPN.

![Fig. 3](image)
There are several white-yellow papules, that are 3 to 5-mm in diameter on the left side of the neck.

![Fig. 4](image)
Arrowheads: In a relatively circumscribed area, some thickened collagen bundles are located in the subpapillary to mid-dermis (H & E stain).

![Fig. 5](image)
Slight paucity of the elastic tissue in the papillary dermis was observed (orcein stain).
DISCUSSION

Cutaneous aging is a gradual process caused by extrinsic and intrinsic factors. Extrinsic aging refers to cumulative environmental damage, mainly ultraviolet light radiation (photoaging). Intrinsic aging results from genetically programmed senescence (chronological aging). Temporary wrinkles were first described as a presentation of intrinsic aging. Another two clinicopathological entities, PXE-PDE and WFPN, have also been regarded as the clinical manifestations of intrinsic skin aging.

PXE-PDE is a distinct entity first described by Rongioletti and Rebora in 1992. Since then, eight cases have been reported. The disease is characterized by whitish-yellow papules resembling PXE on the neck and supraclavicular regions of the elderly and an histological loss of or decrease in the number of elastic fibers in the papillary dermis. Laboratory tests and the usual diagnostic procedures for PXE, including ophthalmologic and cardiovascular studies, are normal. Because of the histological similarity to chronologically aged skin aging, PXE-PDE has been viewed as a manifestation of intrinsic skin aging. The main feature of the elastic tissue in intrinsic skin aging is a loss of the elastic fibers in the papillary dermis. Ultrastructural studies confirmed the absence of elastic fibers in the papillary dermis and revealed elastotic changes as consisting of immature elastic fibers in the subpapillary and mid-dermis. There were also activated fibroblasts with dilated cisternae in their rough endoplasmic reticulum. The elastotic change in PXE-PDE may be related to the initial elastolytic change, and may therefore reflect the process of repairing the tissue damage.

WFPN was described in 1985 by Shimizu and Nishikawa. It has never been observed before 39 years of age (mean age 67). Clinically, multiple asymptomatic, 2 to 3-mm wide, round to oval, non-confluent, non-pedunculated, firm papules are located on the posterior or lateral sides of the neck in elderly people. The papules are not related to hair follicles and range in number from a few to a hundred. There is no associated systemic diseases. Histologically, there is only a slight, focal increase and thickening of collagen fibers in the papillary to mid-dermis under the H & E stain. With special stains of elastic fibers, Shimazu, et al., showed that the amount of elas-

### Table I. The age-related fibroelastolytic syndromes: main differences between PXE-PDE and WFPN

<table>
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<th>Disorders</th>
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<th>Pathology</th>
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<tr>
<td>PXE-PDE</td>
<td>Yellow soft papules coalescent in cobblestone plaques on lateral sides of the neck and supraclavicular areas</td>
<td>Elastic fibers loss in the papillary dermis and reduced in the reticular dermis</td>
<td>Immature elastic fibers and activated fibroblasts in the upper dermis</td>
</tr>
<tr>
<td>WFPN</td>
<td>White non-confluent firm papules on posterior and lateral sides of the neck</td>
<td>Focal thickening of collagen bundles in upper dermis and occasional focal reduction or loss of elastic fibers in upper and reticular dermis</td>
<td>Increase in length of collagen fibers and activated fibroblasts</td>
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EM: electromicroscopic
tic fibers reduced in 10 of their original 25 patients without any morphological abnormalities. Paucity of the elastic fibers was also observed in a few patients, particularly in the papillary dermis. Ultrastructurally, no abnormalities of the elastic fibers were observed. Collagen fibrils seemed tightly compacted with a slight increase in their diameter. Several fibroblasts have abundant rough endoplasmic reticulum.

Moreover, Balus, et al., reported 20 cases and reviewed many of the reports in the literature. They found a considerable overlap between the clinical and histological features of the two entities. Almost all cases of WFPN or PXE-PDE consist of papular eruptions on the neck in elderly people. The lesion color varies and the papules may be isolated or confluent. Histologically, the papules may be characterized by elastolysis, fibrosis or both. PXE-PDE and WFPN are now considered to be two variants of a single disorder that can be more precisely defined as "fibroelastolytic papulosis of the neck" (FEPN) and which appears to be a manifestation of intrinsic skin aging (Table I). FEPN must be differentiated from other papular eruptions that can appear on the neck, including PXE, acrochordons, fibrofolliculoma, anetoderma, post-inflammatory scars, eruptive collagenoma, and perifollicular elastolysis. PXE, which is often clinically indistinguishable from FEPN, can be excluded by the absence of associated cardiovascular and / or ocular changes and the lack of elastic fiber fragmentation with calcium deposition. FEPN lesions are not pedunculated or pigmented, as in the case of acrochordons. The presentation of fibrofolliculoma is most common in the third decade of life and patients with multiple fibrofolliculomas may also have multiple tricodiscomas and acrochordons (Birt-Hogg-Dube syndrome). Mononuclear inflammatory cell infiltrates have been found in all types of anetoderma, regardless of whether the lesions were thought to be clinically inflamed (Jadasson-Pellizari type) or not (Schweninger-Buzzi type). Clinically, moreover, anetoderma is an atrophic, circumscribed, sack-like areas of the slack skin that gives the sensation of a loss of substance rather than firm papules. Although post-inflammatory scarring and WFPN share similar histological changes, one can easily identify the inflammatory stage of the former. The acquired eruptive variant of connective tissue nevi occurs most commonly on the back and the extremities, and it is quite rare for the nevi to be confined to the neck. In perifollicular elastolysis, the papules are located mainly on the trunk and are preceded by acne. The elastic tissue is lost only around the hair follicles.

In conclusion, PXE-PDE and WFPN are two acquired, non-inflammatory fibroelastolytic disorders of the elderly. Although the role of extrinsic factors such as sun exposure cannot be definitively excluded, given their location, the late onset and microscopic and ultrastructural features of both PXE-PDE and WFPN indicate them to be clinical variants of a single disorder, designated FEPN of the same process, namely "intrinsic aging". Considering the asymptomatic and slowly progressive nature of this condition, it is likely that its true incidence has been underestimated.

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