

Fibroelastolytic Papulosis of the Neck

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Fibroelastolytic papulosis of the neck (FEPN) was introduced by Balus et al in 1997. Before this term was used, white fibrous papulosis of the neck (WFPN) and pseudoxanthoma elasticum-like papillary dermal elastolysis (PDE) had differently been used. These two disease entities had clinical similarities such as asymptomatic, white to yellow discolored, papular eruptions on the neck in elderly persons. As names implicated, however, they showed variable histological findings, for examples, fibrosis in WFPN or elastolysis in PDE or both in FEPN. Taken into account together, instead of using WFPN or PDE separately, FEPN might be preferred to describe these two kinds of skin diseases. We report a case of a 36-year-old male patient with papules compatible with FEPN, whose biopsy mainly showed elastolysis.

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The term 'white fibrous papulosis of the neck (WFPN)' was first offered by Shimizu, et al. in 1989, in which the lesions were asymptomatic, whitish papules developed on the neck¹. Histologically, fibrosis of the papillary and upper dermis was characteristic in WFPN and sometimes mild alterations of elastic fiber were accompanied. Meanwhile, clinically similar conditions were reported by other authors with acquired elastolysis of the papillary dermis simulating pseudoxanthoma elasticum (PDE)². Balus et al. recently reviewed the 20 cases of WFPN and PDE, reaching the conclusion that these could be variants of a single disorder described as fibroelastolytic papulosis of the neck (FEPN)³. We herein present a male patient who had skin lesions consistent with FEPN.

CASE REPORT

An otherwise healthy 36-year-old Korean man

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was presented for evaluation of lesions on his neck. He stated that all lesions were asymptomatic and the lesions had been present for 2 to 3 years. On physical examination, there were 2 to 4 mm, round to oval, clearly marginated, ivory-colored, non-follicular, discrete papules over both sides of the neck with a few skin tags located amid the lesions (Fig. 1). His medical, family, and social histories were unremarkable with regard to the development of the lesions. He had not taken any medications. Other than gall bladder polyp in ultrasonogram, there were no abnormalities in routine laboratory work-up including complete blood cell count, urinalysis, blood chemistry, stool examination, Chest PA, tests for hepatitis and human immunodeficiency virus, electrocardiogram, and gastric endoscopy.

The skin biopsy specimen was obtained from the lesion on the right side of the neck.

A biopsy with H & E staining appeared to demonstrate that collagen bundles in papillary and mid-dermis were seemingly normal or slightly increased (Fig. 2). But, the extent of fibrosis or the evidence of elastolysis was not detectable with H&E staining. Masson-trichrome stain and Verhoeff-van Gieson stain showed a focal normal to

Fig. 1. Discrete, round to oval, ivory-colored, 2-4 mm in size, papules on the side of the neck.

Fig. 2. normal to minimal increase in collagen bundles in upper and mid-dermis (H&E stain, $\times 100$).

Fig. 3. focal slight increase in collagen fibers in upper and mid-dermis (Masson-trichrome stain, $\times 400$).

Fig. 4. Marked decrease in elastic fibers in upper and mid-dermis (Verhoeff-van Gieson stain, $\times 200$).

slight increase in the collagen fibers(Fig. 3) and a significant decrease in the elastic fibers(Fig. 4).

DISCUSSION

Clinically, the lesions of WFPN were isolated and whitish papules^{1,3,9}, while those of PDE were coalescent and yellowish papules^{2,10-14}. Histologically, WFPN showed superficial dermal fibrosis with minimal changes in elastic fibers^{3,9}, whereas PDE showed papillary elastolysis without fibrosis¹⁰⁻¹⁵. Some cases posed both of the features^{4,6,13}. Although several authors described WFPN and PDE as separate disease entities, many cases may considerably overlap. Perrin et al. addressed the case clearly demonstrating the histological continuum between WFPN and PDE¹³. Moreover, most cases of WFPN and PDE consist of papulosis which is con-

fined to the neck in elderly people. Balus et al. acclaimed that WFPN and PDE may represent variants of a single disorder designated FEPN³. The lesions tend to develop gradually with no spontaneous regression and are asymptomatic in all cases. The age of onset is generally from late middle to old age, at least more than 40 years old. In our case, relatively young, 36-year-old male patient was involved. The lesions were usually multiple, small, firm, round, elevated, whitish-ivory to slightly yellowish, monomorphous papular eruptions of isolated or confluent in plaques. They were distributed on the nape and/or sides of the neck, occasionally extending up to the border of the scalp or to the shoulders or to the axillae⁹.

Because most of patients with FEPN were elderly people, FEPN could be considered as an aging-related disorder, intrinsic rather than extrinsic^{1,4,5,7}.

To date, there has been no case except this one, reporting the patient with FEPN under the age of 40. There could be a discrepancy about the gender: the European and Middle-Eastern patients were all women^{3,14}, in contrast those reported from Japan were predominantly men¹. Likewise, our patient was a man.

The most noteworthy histological finding was either dermal fibrosis or elastolysis or both, involving the papillary and upper reticular dermis^{3,4,6,13}, which rather rationalize the term fibroelastolysis. However, H&E staining revealed a variation in the appearance of the fibrotic tissues from edematous to sclerodermatous condition. In our case, normal to minimal collagenosis and significant elastolysis were seen. Ultrastructural studies showed numerous large bundles of collagen fibers and elastotic degeneration with clumps of elastin³.

FEPN should be differentiated from other papular eruptions developing on the neck, for instances, acrochordons, fibrofolliculomas and trichodiscomas (Birt-Hogg-Dube syndrome), actinic elastotic disorders, guttate morphea, and pseudoxanthoma elasticum³. Fortunately, histological features of clinically mimicking diseases are distinct from those of FEPN. Guttate morphea can be excluded by abundant fibrosis from mid- to deep dermis with patchy mononuclear cell infiltrate, and pseudoxanthoma elasticum by absence of elastic fiber fragmentation with calcium deposition³. When the cutaneous lesions histologically simulating FEPN develop on the hands, they can be called collagenous and elastotic plaques of the hands¹⁵.

We intended to have our patient undergo laser therapy or cryotherapy for ameliorating the lesions. But, he refused to be treated.

Depending on which aspect can be emphasized, FEPN may show a wide scope of histological features such as fibrosis or elastolysis or both. We diagnosed our case as FEPN with a tendency of elastolysis. In addition, further investigation over complicated histogenesis of FEPN will be needed.

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