Asymptomatic Papules on the Neck

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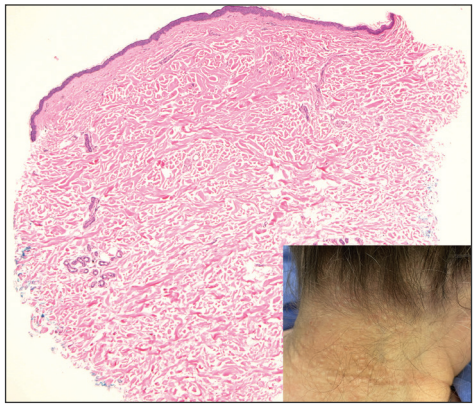
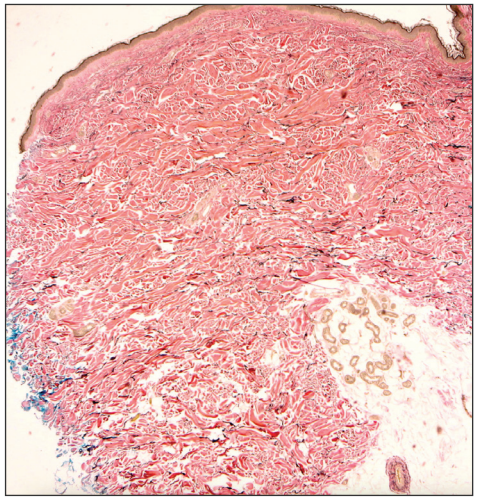


H&E, original magnification ×4 (inset, asymptomatic, 2- to 3-mm, white to flesh-colored papules concentrated on the posterior neck).



Verhoeff-Van Gieson, original magnification ×4.

A 70-year-old woman with a history of osteoporosis and breast cancer presented for evaluation of asymptomatic, 2- to 3-mm, white to flesh-colored papules concentrated on the inferior occipital scalp and posterior neck (inset) for at least several months. She had no additional systemic signs or symptoms, and there was no family history of similar skin findings. A punch biopsy was performed.

THE BEST **DIAGNOSIS IS:**

a. actinic elastosis
b. elastofibroma dorsi
c. fibrofolliculomas
d. pseudoxanthoma elasticum
e. white fibrous papulosis

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THE **DIAGNOSIS:** White Fibrous Papulosis

Given the histopathology findings, location on a sun-exposed site, lack of any additional systemic signs or symptoms, and no family history of similar lesions to suggest an underlying genetic condition, a diagnosis of white fibrous papulosis (WFP) was made. White fibrous papulosis is a relatively rare cutaneous disorder that was first reported by Shimizu et al[1] in 1985. It is characterized by numerous grouped, 2- to 3-mm, white to flesh-colored papules that in most cases are confined to the neck in middle-aged to elderly individuals; however, cases involving the upper trunk and axillae also have been reported.[1-3] The etiology of this condition is unclear but is thought to be related to aging and chronic exposure to UV light. Although treatment is not required, various modalities including tretinoin, excision, and laser therapy have been trialed with varying success.[2,4] Our patient elected not to proceed with treatment.

Histologically, WFP may manifest similarly to connective tissue nevi; the overall architecture is nonspecific with focally thickened collagen and often elastic fibers that may be normal to reduced and/or fragmented, as well as an overall decrease in superficial dermal elastic tissue.[3,5] Therefore, the differential diagnosis may include connective tissue nevi and require clinical correlation to make a correct diagnosis.

Pseudoxanthoma elasticum (PXE) is an autosomal-recessive disorder most commonly related to mutations in the ATP binding cassette subfamily C member 6 (*ABCC6*) gene that tends to manifest clinically on the neck and flexural extremities.[6] This disease affects elastic fibers, which may become calcified over time. Pseudoxanthoma elasticum is associated with ocular complications relating to the Bruch membrane of the retina and angioid streaks; choroidal neovascularization involving the damaged Bruch membrane and episodes of acute retinopathy may result in vision loss in later stages of the disease.[7] Involvement of the elastic laminae of arteries can be associated with cardiovascular and cerebrovascular complications such as stroke, coronary artery disease, claudication, and aneurysms. Involvement of the gastrointestinal or genitourinary tracts also may occur and most commonly manifests with bleeding. Pathologic alterations in the elastic fibers of the lungs also have been reported in patients with PXE.[8] Histologically, PXE exhibits increased abnormally clumped and fragmented elastic fibers in the superficial dermis, often with calcification (Figure 1). Pseudo-PXE related to D-penicillamine use often lacks calcification and has a bramble bush appearance.[9]

Fibrofolliculomas may manifest alone or in association with an underlying condition such as Birt-Hogg-Dubé syndrome, in which lesions are most frequently seen scattered on the scalp, face, ears, neck, or upper trunk.[10] This condition is related to a folliculin (*FLCN*) gene germline mutation. Birt-Hogg-Dubé syndrome also may be associated with acrochordons, trichodiscomas, renal cancer, and lung cysts with or without spontaneous pneumothorax.

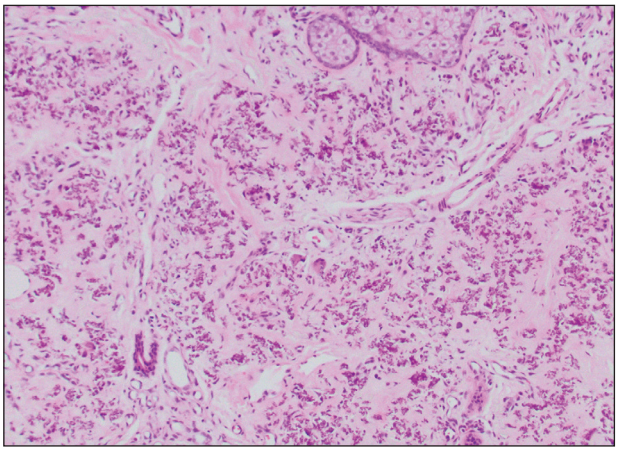

FIGURE 1. Pseudoxanthoma elasticum demonstrates increased abnormally clumped calcified and fragmented elastic fibers (H&E, original magnification ×100).

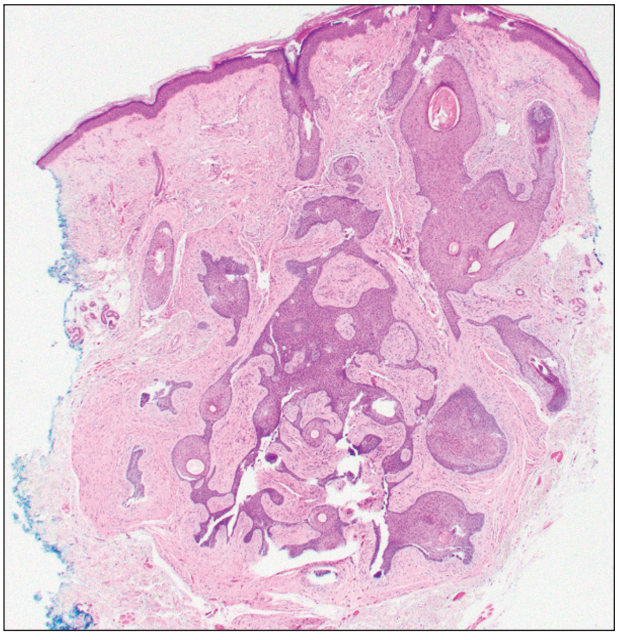

FIGURE 2. Fibrofolliculoma demonstrates epidermal strands originating from a hair follicle associated with prominent surrounding connective tissue (H&E, original magnification ×20).

Less frequently noted findings include oral papules, epidermal cysts, angiofibromas, lipomas/angiolipomas, parotid gland tumors, and thyroid neoplasms. Connective tissue nevi/collagenomas can appear clinically similar to fibrofolliculomas; true connective tissue nevi are reported less commonly in Birt-Hogg-Dubé syndrome.[11]

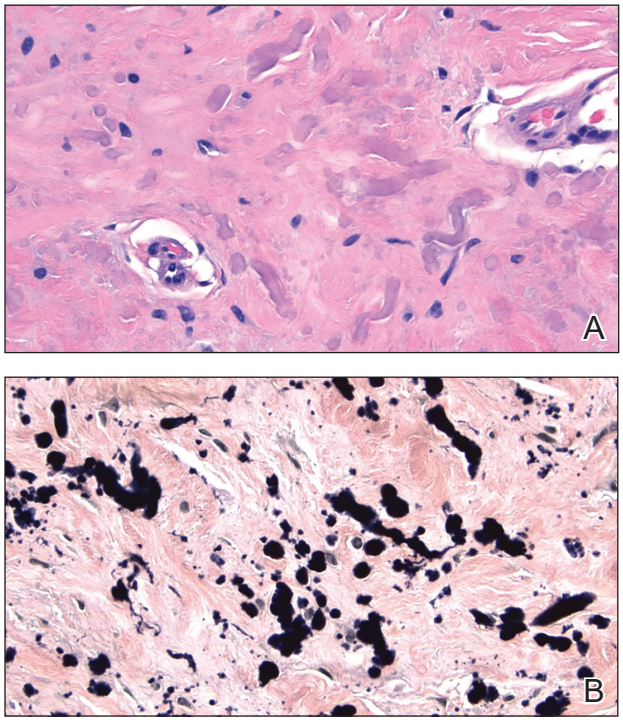

FIGURE 3. A and B, Elastofibroma demonstrates thickened and rounded to beaded elastic fibers (H&E, original magnification ×40), which stain deeply positive with Verhoff-Van Gieson (original magnification ×40).

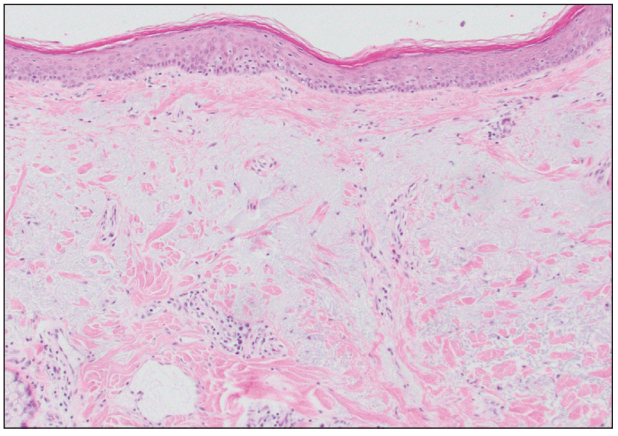

FIGURE 4. Actinic elastosis manifests as basophilic degenerated elastic fibers in the dermis (H&E, original magnification ×100).

Histologically, a fibrofolliculoma manifests with epidermal strands originating from a hair follicle associated with prominent surrounding connective tissue (Figure 2).

Elastofibroma dorsi is a benign tumor of connective tissue that most commonly manifests clinically as a solitary subcutaneous mass on the back near the inferior angle of the scapula; it typically develops below the rhomboid major and latissimus dorsi muscles.[12] The pathogenesis is uncertain, but some patients have reported a family history of the condition or a history of repetitive shoulder movement/trauma prior to onset; the mass may be asymptomatic or associated with pain and/or swelling. Those affected tend to be older than 50 years.[13] Histologically, thickened and rounded to beaded elastic fibers are seen admixed with collagen (Figure 3).

Actinic (solar) elastosis frequently is encountered in many skin biopsies and is caused by chronic photodamage. More hypertrophic variants, such as papular or nodular solar elastosis, may clinically manifest similarly to WFP.[14] Histologically, actinic elastosis manifests as a considerable increase in elastic tissue in the papillary and superficial reticular dermis (Figure 4).

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