Case Letter

Papular Elastorrhexis

To the Editor:

Papular elastorrhexis (PE) is an elastic tissue disease that presents with white asymptomatic papular eruptions that are histologically characterized by loss and fragmentation of dermal elastic fibers. Papules usually are white; measure 1 to 5 mm in diameter; and appear in a scattered pattern on the abdomen, chest, back, shoulders, and upper extremities. Patients generally do not have a history of infection, acne, or trauma at the lesion site.[1] Papular elastorrhexis was first described by Bordas et al[2] in 1987 as a variant of nevus anelasticus. Since then, approximately 25 cases have been reported in the English-language literature, according to a PubMed search of articles indexed for MEDLINE using the term *papular elastorrhexis*.[3-6] The majority of cases involved women in the second decade of life. Papular elastorrhexis patients only have cutaneous symptoms, and the disease's etiology and pathogenesis remain unknown.[1,6]

A 26-year-old woman presented to our polyclinic with eruptions over the lower portion of her chest of 1 year's duration without any preceding signs or symptoms. She did not have a family history of similar lesions. Dermatologic examination showed multiple hypopigmented papular lesions that measured 1 to 5 mm in diameter with smooth surfaces (Figure 1). The patient did not report a history of trauma, acne, or infection at the lesion site. Routine laboratory analysis did not reveal any abnormalities. Histopathologic examination of the lesional skin showed diminution and fragmentation of dermal elastic fibers (Figure 2). Direct radiography showed no osseous pathology. Based on these findings, the patient was diagnosed with PE.

Papular elastorrhexis is an uncommon nevus of connective tissue characterized by a decrease, loss, and fragmentation of elastic fibers.[1,7] Papular elastorrhexis is thought to be a variant of Buschke-Ollendorf (BO) syndrome by some authors,[7,8] whereas others consider it a different entity.[4,9] Buschke-Ollendorf syndrome is inherited in an autosomal-dominant fashion and is characterized by widespread white papules and osteopoikilosis. Although it is thought that PE is not genetically inherited, a report of a mother and her 2 children with PE suggests that it may be a genetically inherited variant.[8] Although PE lesions are clinically similar to those of BO syndrome, PE differs by its absence of familial inheritance, late onset, and absence of signs of osteopoikilosis.[7,9]

Histopathologic examination of PE patients shows decrease, loss, and fragmentation of elastic fibers, whereas accumulation of elastic fibers that form nets of bundles are observed in BO syndrome.[7,8] Based on the age at onset of symptoms, absence of familial history and osteopoikilosis in the bones, and histopathologic findings of diminution and fragmentation of the elastic fibers, our patient was diagnosed with PE.

Other diseases that should be included in the differential diagnosis of PE are nevus anelasticus,

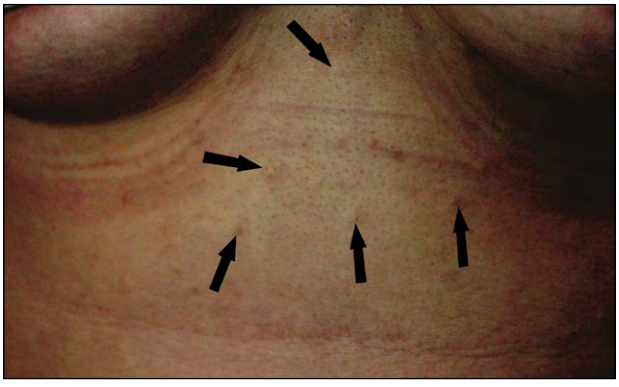

Figure 1. Multiple hypopigmented papular lesions (arrows) on the inferior chest.

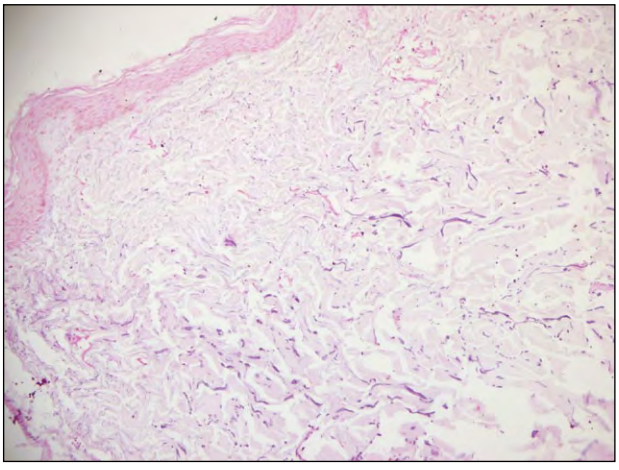

Figure 2. Distinct loss, thinning, and fragmentation of elastic fibers in the papillary dermis (van Gieson, original magnification ×20).

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eruptive collagenoma, papular acne scars, anetoderma, and mid dermal elastolysis. Nevus anelasticus is characterized by perifollicular papules that form bundles on the trunk, whereas PE papules are not perifollicular. Fiber loss and degeneration in the upper and mid dermis also are present in nevus anelasticus.[1,6] Eruptive collagenomas exhibit loss of elastic fibers in addition to thickened and homogenized collagen bands. Papular acne scars with follicular localization are associated with a history of acne and do not occur on the abdomen and thighs. In anetoderma and mid dermal elastolysis, elastolysis occurs without elastorrhexis, and collagen appears normal.[6]

There is no known treatment of PE. In 1 case, papules were flattened by 20 mg/mL of intralesional triamcinolone acetonide.[10] Our patient was treated with 10 mg/mL of intralesional triamcinolone acetonide twice at intervals of 3 weeks; however, no improvement in her skin lesions was observed at the end of the 6 weeks.

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