Papular Elastorrhexis

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Papular elastorrhexis (PE) is a rare connective tissue disease, histopathologically characterized as decreased and fragmented elastic fibers in the reticular dermis, with or without change to the collagen bundles. It presents as small, white, nonfollicular papules predominantly scattered over the chest, shoulders, or back, and is most often seen in females of the second decade. Although all reported cases of PE have been asymptomatic, a 21-year-old woman presented with a one-year history of multiple, small skin-colored papules on the back which were associated with an intermittent itching sensation. Histopathologic examination revealed fragmented elastic fibers with focal homogenization of collagen in the reticular dermis. Therefore, we report a rare case of papular elastorrhexis with "pruritus".
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Key Words: Papular elastorrhexis, Elastic fiber

INTRODUCTION

Papular elastorrhexis (PE) was first described in 1987 by Bordas1 as multiple, discrete, nonfollicular, 1 to 5 mm diameter-sized white papules evenly distributed over the back, chest, shoulders, and upper extremities. It usually occurs in females, first appearing during the second decade of life. It histologically characterizes as decreased and fragmented elastic fibers, with or without changes to the collagen bundles in the reticular dermis.

Since 1987 there have been reports of at least 17 cases of PE in the English literature[1-5], and almost all of the cases had no symptoms. However we describe a patient with pruritic papules on the back which showed the typical histologic finding of PE.

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CASE REPORT

A healthy 21-year-old woman presented with a 1-year history of multiple skin-colored papules on the upper back. The history of trauma to the lesions was unremarkable, except for excoriation due to an itching sensation. However, the papules had already been present several months before excoriation. She had no past history of acne and no family history of similar lesions or other skin diseases. The physical examination revealed multiple, white-

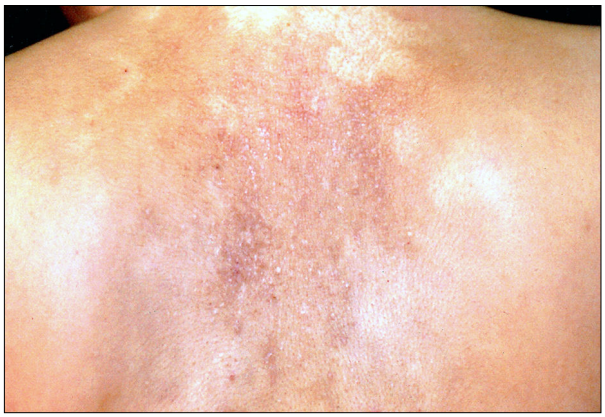

Fig. 1. Multiple, nonfollicular, white papules on the back.

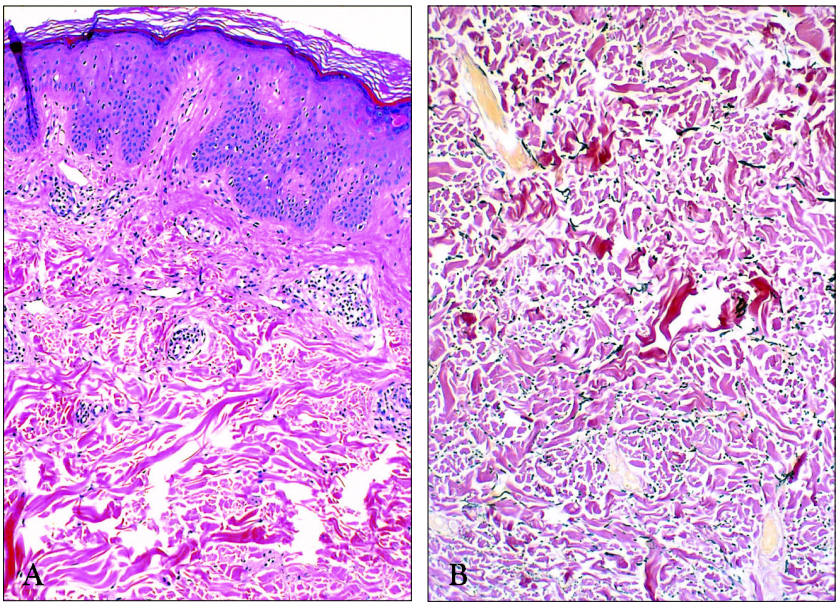

Fig. 2. (A) Focal area of thickened, homogenized collagen bundles throughout the dermis (H&E, × 100). (B) Fragmentation of elastic fibers throughout the dermis (Elastic stain, × 100).

colored papules dominantly on the upper back (Fig. 1). The lesions were nonfollicular and did not coalesce to form plaques.

A punch biopsy from a papule lesion revealed a focal area of thickened and homogenized collagen bundles in the reticular dermis, and perivascular lymphohistiocytic infiltration (Fig. 2A). Elastic tissue staining showed elastic fibers of the reticular dermis were intensively fragmented which resulted in a speckled appearance (Fig. 2B), but the perifollicular area do not show any changes. Congo- red staining revealed no amyloid deposition in the lesion. On the basis of the clinical and histopathologic findings, she was diagnosed as having PE.

DISCUSSION

Papular elastorrhexis (PE) is a rare condition, characterized by asymptomatic papules and fragmentation of dermal elastic fibers. In 1987 Bordas et al.[1] described the first case of papular elastorrhexis which consisted of several small yellowish papules on the trunk. It was thought to be a variant of nevus anelasticus due to the reduction and fragmentation of elastic tissue. Sears et al.[2] suggested that this disorder represented a variant of a connective tissue nevi. Schirren et al.[6] suggested that PE was a mild form of Buschke-Ollendorff syndrome because familiar PE had been described. In 2002, Choonhakarn et al.[4] described the distinct and repeatable features that are seen in reported cases of PE, and proposed that PE is a distinct variant of elastic tissue nevi, not an incomplete form of Buschke-Ollendorff syndrome. However, recently there has been no convincing data to support the theory that PE is a variant of connective tissue nevus. Therefore PE appears to be a distinct entity that occurs in adolescence as tiny, white, asymptomatic papules scattered over the torso, with no predilection of the follicular areas and no tendency to group into plaques. In histopathologic examination, the papules demonstrate decreased and fragmented elastic fibers within the reticular dermis. Collagen bundles can be thickened and homogenized, or normal. Some of the cases show a perivascular infiltrate of lymphocytes and histiocytes in the superficial and deep dermis[3].

Because of the decrease and alterations in the elastic fibers, several connective tissue nevus variants or secondary scarring must be considered in the differential diagnosis of PE. Postacne scars or secondary scarring may appear clinically similar but are usually characterized by follicular papules with a marked decrease of elastic fibers surrounding hair

follicles and a laminated arrangement of collagen, whereas PE is classically characterized by nonfollicular papules with elastorrhexis, not elastolysis. Moreover our case had no history of prior trauma or acne. Eruptive collagenoma consists of multiple, asymptomatic papules that histologically shows a typically thickening and homogenization of collagen fibers, and a decrease or degenerative change of elastic fibers that seems to be secondary to increasing collagen fiber. Nevus anelasticus is a congenital disorder with follicular papules that are grouped into patches. Although this disorder shows a decrease in the elastic fibers without changes of collagen in the lesion, it differs significantly from PE, clinically. PE appears in adolescence and papules tend to be nonfollicular, evenly scattered over the affected areas, and do not tend to coalesce.

The paucity of reports on PE and the limited details provided in several of these make it impossible to determine whether PE could be associated with pruritus. However, other probable cutaneous conditions were ruled out and the significant factor provoking pruritus was not found. We describe a case of "PE with itching sensation" which presented clinically as typical, multiple, white nonfollicular papules on the back, and histologically as fragmented elastic fibers in the reticular dermis which was consistent with papular elastorrhexis.

REFERENCES

1. Bordas X, Ferrandiz C, Ribera M, Galofre E. Papular elastorrhexis: A variety of nevus anelasticus. Arch Dermatol 1987;123:433-434.
2. Sears J, Seabury Stone M, Argenyi Z. Papular elastorrhexis: A variant of connective tissue nevus. J Am Acad Dermatol 1988;19:409-414.
3. Lewis KG, Bercovitch L, Dill SW, et al. Acquired disorders of elastic tissue: Part II. decreased elastic tissue. J Am Acad Dermatol 2004;51:165-185.
4. Choonhakarn C, Jirarattanapochai K. Papular elastorrhexis: a distinct variant of connective tissue nevi or an incomplete form of Buschke-Ollendorff syndrome. Clin Exp Dermatol 2002;27:454-457.
5. Buechner SA, Itin P. Papular elastorrhexis. report of five cases. Dermatology 2002;205:198-200.
6. Schirren H, Schirren CG, Stolz W, et al. Papular elastorrhexis: A variant of dermatofibrosis lenticularis disseminata (Buschke-Ollendorff syndrome). Dermatology 1994;189:368-372.