

## A Case of Papular Elastorrhexis

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*Papular elastorrhexis is a rare disease, characterized by multiple nonfollicular white papules that usually occur on the trunk. Papular elastorrhexis occurs during childhood and adolescence. Histopathologically, the elastic fibers are decreased and they may appear in thin and fragmented forms. Herein, we report a 20-year-old Korean woman with asymptomatic multiple nonfollicular white firm papules that were scattered over the anterior chest. (Ann Dermatol (Seoul) 19(4) 185~188, 2007)*

Key Words: Elastic fiber, Papular elastorrhexis

### INTRODUCTION

Connective tissue nevi are dermal connective tissue hamartomas that are characterized by an imbalance in the relative amount and distribution of collagen, elastin or proteoglycan<sup>1</sup>. If elastic fibers are exclusively involved, then elastic tissue nevus would be the appropriate term. Papular elastorrhexis is an uncommon disease that usually occurs in childhood or adolescence with multiple, discrete, white, papules, which are located mostly on the trunk.

Bordas et al.<sup>2</sup> first used the term papular elastorrhexis in 1987 to describe acquired, multiple, 2 to 5 mm, flat, firm, painless, oval-shaped papules. To the best of our knowledge, there have been only 18 reported cases of papular elastorrhexis in literature<sup>2-10</sup>. Herein, we report a case of 20-year-old-Korean woman who had asymptomatic multiple, discrete, firm, palpable white papules on the anterior chest for several months; We also include a brief literature review.

### CASE REPORT

A 20-year-old Korean woman presented with

several months, history of asymptomatic, multiple papules on the anterior chest, which had been gradually increasing in number. On physical examination, we observed multiple discrete nonfollicular 2 to 5 mm sized firm, slightly indurated, whitish papules that were scattered symmetrically on the anterior chest (Fig. 1). There was no history of bleeding or any change in the morphology of the skin lesions since their first appearance. She denied a history of preceding acne or other inflammatory skin disorders on the involved area. There was no family history of similar lesions or other skin diseases. No bony abnormality was found in the long bones and pelvis on radiologic examination. A biopsy specimen from a lesion on the anterior chest showed normal epidermis with focal areas of

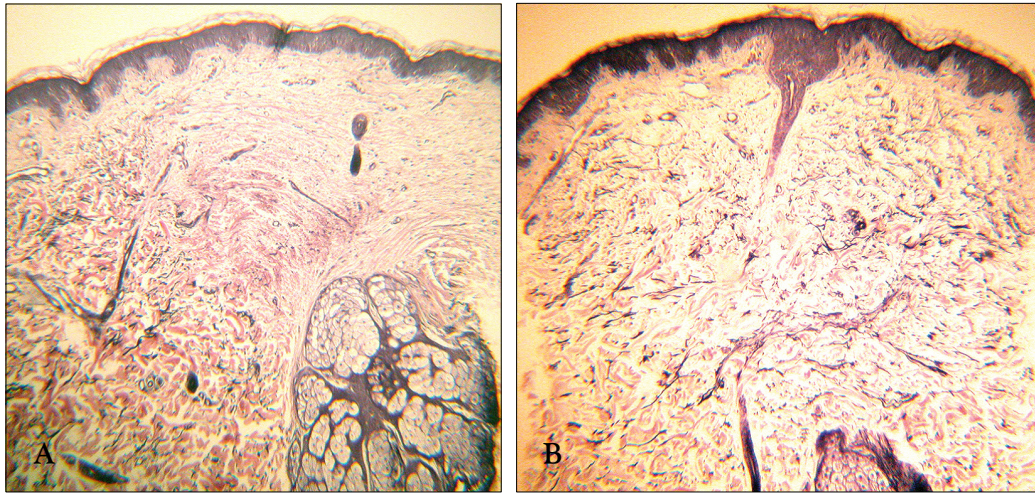


**Fig. 1.** Asymptomatic multiple discrete firm nonfollicular whitish papules on the trunk (Inset: close-up view).

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**Fig. 2.** (A) Focally decreased and fragmented elastic fibers in the upper dermis of the lesion, and (B) in the normal skin (Verhoeff-van Gieson stain,  $\times 100$ ).

homogenized and condensed collagen bundles in the upper dermis. Verhoeff-van Gieson stain revealed decreased and fragmented elastic fibers in the upper dermis (Fig. 2). The lesions persisted without any treatment.

## DISCUSSION

Papular elastorrhexis occurs predominantly in the 2nd decade as discrete whitish firm papules with the focal decrease and fragmentation of elastic fibers.

We reviewed the clinical and histological features of 18 cases of papular elastorrhexis since the first report by Bordas *et al.*<sup>2</sup> in 1987 (Table 1)<sup>2-10</sup>. The average age of onset was 11.9 years with a range from 3 to 18 years. Ten of 18 patients were girls and the trunk and upper extremities were the most common locations. The descriptions about patterns of collagen were more varied; 2 cases had normal collagen, 13 cases had homogenized and condensed collagen bundles and one case had coarse collagen.

The exact origin of papular elastorrhexis is still unknown; however, it has been debated whether papular elastorrhexis is a single entity or an abortive form of dermatofibrosis lenticularis disseminate (DLD, Buschke-Ollendorff syndrome)<sup>4</sup>. DLD is a rare, autosomally dominant inherited syndrome characterized by yellow or flesh-colored papules, nodules and plaques<sup>11</sup>. The cutaneous lesions of DLD are best classified as connective tissue nevi of

the elastic type and histologically, the lesions are characterized by excessive amounts of unusually broad, interlacing elastic fibers in the dermis without fragmentation of the fibers. The skin lesions are usually associated with osteopoikilosis; this consists of multiple circumscribed opacities at the ends of long bones. However, in this case, we could not find any bony abnormalities including osteopoikilosis or similar skin lesions in her family. Histopathologically, instead of accumulation of thick, branching elastic fibers, decreased and altered elastic fibers were observed in our case.

Wilson *et al.*<sup>12</sup> reported that papular acne scars represent postacne scars and they are indistinguishable from those described as papular elastorrhexis. However, papular elastorrhexis differs from postacne scars in some aspects. First of all, papular elastorrhexis is nonfollicular oriented. In addition, acne lesions are not usually located on the arms, thighs and abdomen. These findings are not consistent with postacne papular scars. Moreover, the histological features of acne scars such as flattening of the rete ridge, laminated arrangement of collagen and altered vasculature can not be found in papular elastorrhexis. In our case, the patient denied any previous history of acne and the lesions were not located in the follicular lesions. Furthermore, the histopathologic findings showed decreased and fragmented elastic fibers in the upper dermis, rather than displaying the features of acne scars.

Eruptive collagenoma, which is another variant of

**Table 1.** Summary of the clinical and histopathological findings in the reported cases of papular elastorrhexis

Case No	Age/Sex	Duration	Site	Elastic fiber	Collagen bundles	Follicles	Treatment
Bordas et al <sup>2</sup>	17/M	3 yrs	Abd, chest, back	D	NS	Nonfollicular	NS
Sears et al. <sup>3</sup>	21/F	7 yrs	Back, deltoid region	D, F	H	Nonfollicular	None
Sears et al <sup>3</sup>	21/F	5 wks	Chest	D, F	H, C	Nonfollicular	None
Schirren et al <sup>4</sup>	17/F	Few yrs	Back, UE, LE	D, F	Normal	Nonfollicular	NS
Schirren et al <sup>4</sup>	20/M	NS	Trunk, UE, LE	D, F	Normal	Nonfollicular	NS
Schirren et al <sup>4</sup>	45/F	4 yrs	Trunk, LE	NS	NS	Nonfollicular	NS
Lee et al <sup>5</sup>	6/M	1 mon	Trunk, UE	D, F	H	Nonfollicular	None
Lee et al <sup>5</sup>	10/M	5 yrs	Trunk, LE	D, F	H	Nonfollicular	None
Lee et al <sup>5</sup>	7/M	6 mons	Neck, abd	D, F	H	Nonfollicular	None
Lee et al <sup>6</sup>	4/M	1 yr	Chest, back	Focal loss, F	NS	Nonfollicular	ILI
Choonhakarn et al <sup>7</sup>	18/F	4 yrs	Abd, back	D, F	H, C	Nonfollicular	NS
Buechner et al <sup>8</sup>	9/F	1 yr	Abd, back	D, F	T, C	Nonfollicular	NS
Buechner et al <sup>8</sup>	12/F	4 mons	Trunk	D, F	T, C	NS	NS
Buechner et al <sup>8</sup>	12/F	6 mons	Trunk, UE	D, F	T, C	NS	NS
Buechner et al <sup>8</sup>	11/M	2 yrs	Trunk, LE	D, F	T, C	NS	NS
Buechner et al <sup>8</sup>	11/F	1 yr	Arms, thighs, back	D, F	T, C	NS	NS
Choi et al <sup>9</sup>	19/F	Several mons	Trunk	D, F	H	NS	ILI
Ryder et al <sup>10</sup>	16/F	3 yrs	Back, shoulder	Nearly absent	Coarse	Nonfollicular	NS
Our Case	16/F	Several mons	Ant. chest	D, F	H	Nonfollicular	None

Abd: Abdomen; D: Decreased; NS: Not stated; F: Fragmented; H: Homogenized; C: Condensed; UE: Upper extremities; LE: Lower extremities; ILI: Intralesional injection; T: Thickened

connective tissue nevi disorders, is found as multiple, white or flesh-colored 2-5 mm papules on the trunk occurring before or after adolescence and showing decreased elastic fibers and thickened, homogenized collagen<sup>10</sup>. Various reports have debated about the disease spectrum between eruptive collagenoma and papular elastorrhexis<sup>3,10</sup>. Papular elastorrhexis is known to be more uniformly sized and smaller than eruptive collagenoma but both are found as multiple papules showing de-

creased elastic fibers and homogenized collagen.

There is no reliable treatment for papular elastorrhexis. Improvement after intralesional injection of triamcinolone has been reported in two cases<sup>6,9</sup>. Oral antihistamine, oral and topical isotretinoin and benzoyl peroxide has also been tried, but they were not effective<sup>3</sup>.

Papular elastorrhexis has been only rarely described in literature, the clinical changes of papular elastorrhexis are subtle and they may go

unnoticed in many instances. Recognition of this entity is important to prevent the use of unsuccessful therapeutic agents.

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