A case of papular elastorrhexis

Müzeyyen Gönül[1], Göknur Bilen[1], Aysun Gökce[2], Murat Alper[1]

[1]Dermatology Clinic, Dışkapı Education and Research Hospital, Ankara, Turkey
[2]Pathology Clinic, Dışkapı Education and Research Hospital, Ankara, Turkey

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Papular elastorrhexis (PE) is a rare disorder of elastic tissue characterized by asymptomatic, nonfollicular, whitish or flesh-coloured, monomorphous, discrete, oval to round papules [1–5]. One to five mm-sized papules are symmetrically distributed on the chest, abdomen, back and upper limbs [1, 3, 4]. Some PE cases may be underestimated because of the asymptomatic course of the lesions or misdiagnosed because of rarity of the disorder and similarity of the lesions to acne scars [2]. Up to now, fewer than 30 PE cases have been reported [4].

A 22-year-old man presented with asymptomatic, flesh-coloured papules on the trunk and upper arms. The lesions had first appeared when he was 13–14 years old and slowly progressed over years. He did not define antecedent trauma or local inflammation. He had acne vulgaris history and he had taken isotretinoin therapy for acne vulgaris 2 years before admission. While acne lesions regressed, the papules did not change as a result of this therapy. There were no other significant findings in his personal and family history. Dermatological examination revealed multiple 1–5 mm-sized, flesh-coloured, firm, nonfollicular discrete papules on the upper regions of the chest, back and upper arms (Figure 1). Also, keratosis pilaris was observed over the lateral surface of upper arms. Routine laboratory tests were within normal limits. Histopathological examination of papules showed perivascular mild lymphoid infiltrate in the superficial dermis and mild homogenization of collagen fibres (Figure 2 A). Fragmentation and diminution, even loss in some areas of elastic fibres were seen by Verhoeff-van Gieson staining in histopathological examination (Figure 2 B). Histopathological examination did not show any follicle in or near the lesion. The lesions were diagnosed as PE based on clinical and histopathological findings.

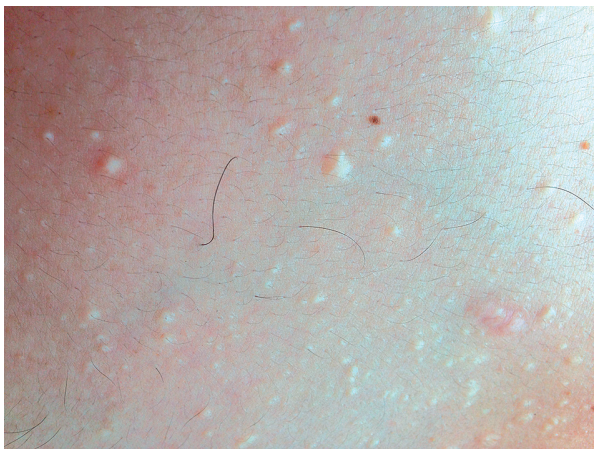

Figure 1. Multiple 1–5 mm-sized, flesh-coloured, firm, discrete papules on the upper chest

Papular elastorrhexis is a rare disorder with no systemic associations and family history [2, 3]. It occurs usually in childhood or adolescence such as in our patient. Most of the reported cases were female [1–4]. Histopathologically, PE displays prominent fragmentation and loss of elastic fibres with or without changes in collagen bundles in the dermis [3].

Differential diagnosis of PE consists of many dermatological entities such as nevus anelasticus, abortive form of Buschke-Ollendorff syndrome, anetoderma, papular acne scars (Table 1). It is controversial whether PE is a distinctive entity or not because of clinical and histopathological similarity to these dermatological entities. Although PE is defined as a distinct variant of connective tissue nevi that are hamartomas characterized by an imbalance in the relative amount and distribution of dermal connective tissue components, the opinion that PE is a separate entity is more accepted today because PE is usually acquired, nonfollicular and sparsely located, and histopathologically has prominent elastic tissue fragmentation [1, 2, 4].

Wilson *et al.* evaluated 133 dermatology outpatient patients and detected small, hypopigmented papules on the upper part of the trunk in 28% of the patients. They found that there was a statistically significant correlation between papules and a history of truncal acne.

Address for correspondence: Müzeyyen Gönül, Dermatology Clinic, Dışkapı Education and Research Hospital, Yıldızevler Mah, 742. Sok, Aykon Park sitesi, A Blok, No: 3/3, Ankara, Turkey, e-mail: muzeyyengonul@gmail.com
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Table 1. Differential diagnosis of PE

Variable	Epidemiology	Clinical features	Histopathology	Associated findings
Papular elastorrhexis	F > M, 2nd decade	Asymptomatic, nonfollicular, 1–5 mm-sized white flesh-coloured firm papules on the trunk and shoulder	More prominent fragmentation and discrete loss of elastic tissue Perivascular lymphocytes and macrophages dermal infiltrate	–
Nevus anelasticus	Few cases reported Congenital > acquired	Asymptomatic, flat, pink-red follicular papules in asymmetric cluster or confluent plaques in the pectoral region	More prominent loss of elastic tissue and moderately fragmentation of elastic fibres Lack of infiltration	–
Buschke-Ollendorff syndrome	Inherited; AD Early age onset > adult onset	DLD: disseminated, skin-coloured, pea-sized papules on the trunk and extremities Elastomas: asymmetric distribution, flesh-coloured, yellowish tightly grouped papules which may coalesce to form plaques	Accumulation of thick, branching elastic fibres	Osteopoikilosis
Anetoderma	F > M, children and adults	Multiple 5- to 25-mm diameter, round, finely wrinkled, atrophic, flaccid, saclike patches	Loss of elastic tissue; histopathologic study and EM may also show fragmentation of elastic fibres in papillary, mid, or deep dermis	Primary type: not preceded by inflammatory dermatosis Secondary type: preceded by inflammatory dermatosis Both may be associated with systemic disorders
Acne scars	Adolescents, adult	Small, asymptomatic, hypopigmented follicular papules on the upper trunk	Perifollicular or parafollicular lesions, attenuated of both elastic and collagen fibres, a mild increase in fibroblasts and small blood vessels	Acne
Pseudoxanthoma elasticum	Inherited F > M, childhood onset	Small, yellowish papules coalescing to plaques on the neck, abdomen and axillae	Accumulation of fragmented and calcified elastic fibres	Angioid streaks, hypertension, angina, claudication

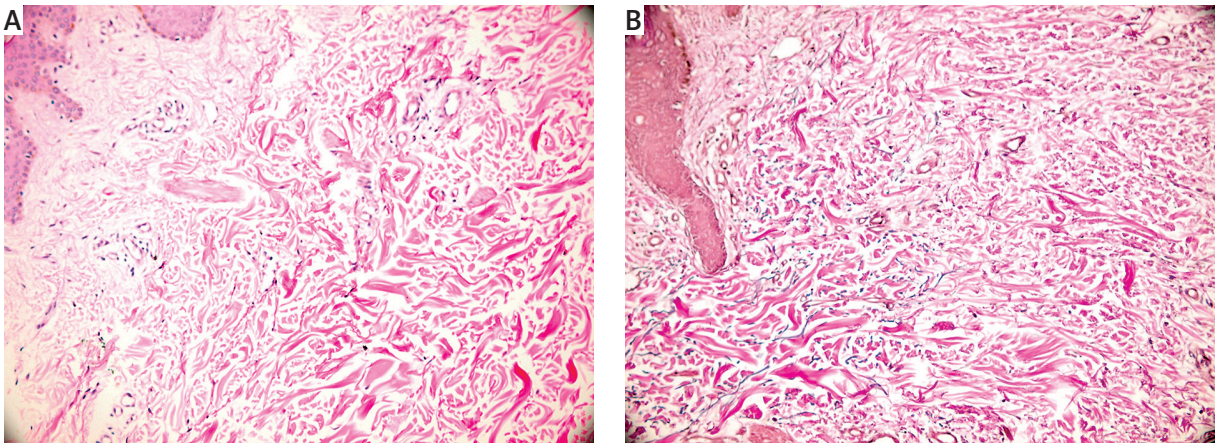

Figure 2. A – Perivascular mild lymphoid infiltrate in the superficial dermis and mild homogenization of collagen fibres (H + E, 200×) **B** – Diminished and fragmented elastic fibres in dermis (Verhoeff-van Gieson, 200×)

Histopathological examination of these papules showed changes consistent with perifollicular scars and these papular lesions were evaluated as post-acne papular scars by Wilson *et al.* [5].

We think that the diagnosis of our case is PE because of the onset age, location, clinical appearance (nonfollicular flesh papules) and histopathological findings (nonfollicular, fragmentation and diminution of elastic fibres) of the papules. Also, we think that PE is a distinct entity from the connective tissue nevi. However, the relation between PE and acne vulgaris is not obvious yet. We think that there may be a possible etiopathological relation between acne and PE because of our case who had a history of acne vulgaris and the data of Wilson *et al.* [5]. Further studies are needed to explain this relation. Our view is that there may be two forms of PE including de novo incipient and subsequent to acne vulgaris or another inflammatory disease.

Whatever the etiopathological factor is, the diagnosis of PE by the dermatologist is important to alleviate the worry of the patient and avoid unnecessary investigations. Dermatologists must keep in mind this unusual entity in patients with flesh-coloured papules and biopsy of the lesions must be performed.

Conflict of interest

The authors declare no conflict of interest.

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