A Case of Papular Elastorrhexis in a Young Female

Dear Editor,

Papular elastorrhexis (PE) is a rare variant of connective tissue nevus. Patients present with multiple non-follicular oval white or yellowish papules on the trunk or limbs; dermal elastic fibers are decreased and fragmented on histology. Although familial cases have been reported, the condition is usually acquired after adolescence and the second decade of life and is more common in women.[1]

A 22-year-old woman presented with multiple asymptomatic skin colored to whitish raised lesions over back, abdomen, neck and proximal aspects of upper limbs which appeared when she was 3 years of age. Lesions had progressed in size and number and had remained stable since the past 6 years and had never resolved completely or partially. There was no family history of similar lesions. There was no history of preceding trauma or any inflammatory disorders like acne in the involved areas. The past medical, surgical, menstrual, and drug history were unremarkable. On examination, multiple, discrete, non-follicular, skin colored to hypopigmented papules were present all over the back, neck, abdomen and both the forearms ranging several millimeters in size [Figures 1 and 2]. There was no tenderness or induration present. Surrounding skin was unremarkable. Hair, nail, mucosae were normal. Systemic examination and routine laboratory tests were normal. Histopathology from one of the lesions on the back, showed an increased number of thin collagen bundles in the upper reticular dermis arranged parallel to the surface with an increased number of thin-walled capillaries many of which were dilated. There was no inflammatory infiltrate or neoplasm. Overlying epidermis was thin and flat. Special stain for elastin, Verhoeff Van Gieson showed reduced and fragmented elastic fibers in the upper dermis [Figures 3 and 4]. On the basis of clinical features and supporting histopathological findings, the diagnosis of PE was made. The patient was prescribed oral isotretinoin 40 mg daily and topical tretinoin gel 0.04% but was lost to follow up.

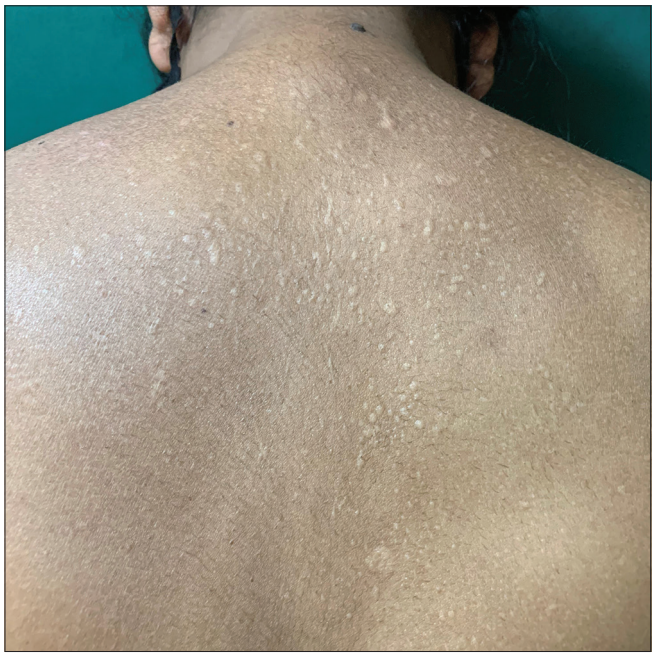

Figure 1: Multiple non-follicular skin colored to white papules distributed on the back

The first case of PE was described by Bordas *et al*.[2] in 1987. It is an elastic tissue disease whose etiology and pathogenesis remain unknown. Some authors consider familial PE as an abortive incomplete variant of Buschke–Ollendorff syndrome (BOS) without associated osteopoikilosis whereas others consider it as a distinct entity from BOS. A recent electron microscopic study in PE highlighted the diminution and degeneration of fibroblasts and elastic tissue as well as swollen collagen bundles, suggesting that the elastic tissue disorder may result from disorganized fibroblasts. Some authors consider PE as a reparative process based on clinical and histopathological findings. The majority of cases occur in women in the first or second decade of life, in an episodic and acquired manner.[3] There is usually no history of trauma, inflammation or previous acne.[3,4] It presents clinically as multiple non-follicular, non-confluent whitish papules, measuring 5 mm or less, distributed mainly over the upper part of the trunk (shoulders and proximal upper arms). The histopathological assessment of the lesions reveals characteristic rarefaction and important fragmentation of the elastic fibers of the reticular dermis.[4,5] There can be a perivascular lymphohistiocytic infiltrate. The collagen is thickened or normal.[3] Dermoscopic features of PE may be unique and include honeycomb-like reticular pigment surrounded by radial pigment, without vessel involvement, however, dermoscopy was not performed in our patient.[6] In postacne scars, there is a history of preceding acne and secondary scarring involves follicular

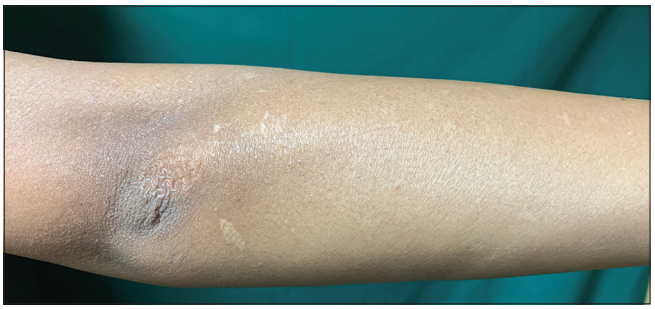

Figure 2: Few non-follicular skin colored papules over the right arm

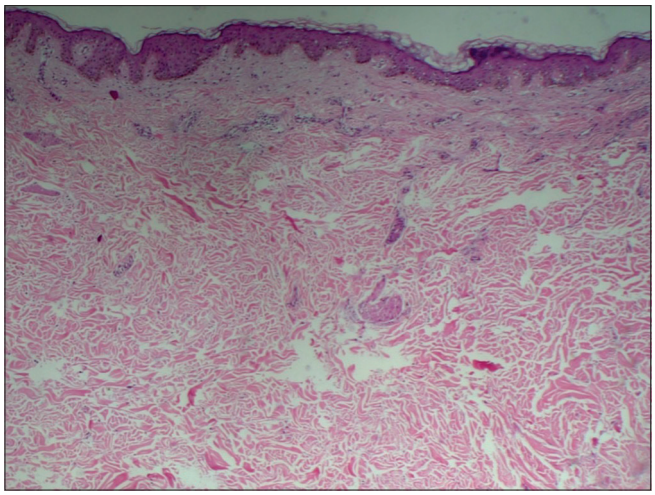

Figure 3: Increased number of thin collagen bundles in upper reticular dermis arranged parallel to the surface, the overlying epidermis is thin and flat. (Hematoxylin and Eosin, ×10)

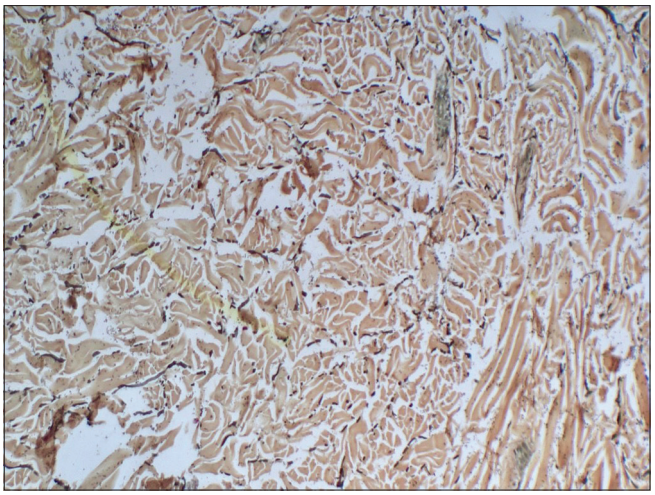

Figure 4: Reduced and fragmented elastic fibers in the upper dermis. (Verhoeff Van Gieson elastic stain, ×10)

papules with a marked decrease of elastic fibers, whereas PE is classically characterized by non-follicular papules with elastorrhexis. In anetoderma, there are atrophic plaques or macules with fine wrinkles and bag like herniation of the underlying tissue. Buttonhole sign is present in anetoderma. There is a loss of elastic fibers, and elastorrhexis may occasionally affect the entire dermis.[7] Nevus anelasticus is usually located on mammary areas as non-symmetrical perifollicular papules and is characterized by prominent loss of elastic fibers rather than fragmentation. Buschke-Ollendroff syndrome is an autosomal dominant disorder with elastomas and dermatofibrosis lenticularis disseminata. Dermatofibrosis lenticularis disseminata shows clinically similar features to PE; however, histopathological study has demonstrated an increase in the number of abnormal collagen fibrils. Eruptive collagenoma almost always shows prominently increased collagen fibers, while PE may or may not show changes of collagen bundles. Mid dermal elastolysis is characterized by patches and plaques of diffuse wrinkled skin with band like loss of elastic fibers in the middermis. Perifollicular elastolysis involves the complete loss of elastic fibers surrounding hair follicles. Pseudoxanthoma elasticum is characterized by coalescing yellow macules and papules with plucked chicken skin, with histology showing mineralized elastic fibers in mid and deep reticular dermis. Cutis laxa is an inherited disorder with pendulous, inelastic skin with an aged facies. After a gradual evolution phase over the years, the lesions of PE are relatively stable, with no tendency for spontaneous resolution. Similar cases of PE were reported by Choi *et al*.[8] in international literature and Xue *et al*.[9] in Indian literature respectively. Currently, the treatment of PE is limited to very few options like topical benzoyl peroxide, topical retinoids and oral isotretinoin; however, the response is variable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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References

1. Schirren H, Schirren CG, Stolz W, Kind P, Plewig G. Papular elastorrhexis: A variant of dermatofibrosis lenticularis disseminata (Buschke-Ollendorff syndrome)? Dermatology 1994;189:368-72.
2. Bordas X, Ferrandiz C, Ribera M, Galofré E. Papular elastorrhexis: A variety of nevus anelasticus? Arch Dermatol

1987;123:433-4.
3. Thomé EP, Steglich RB, Meotti CD, Schwartz J, Boff AL. Case for diagnosis. Papular elastorrhexis. An Bras Dermatol 2012;87:651-53.
4. Emre S, Metin A, Demir-Pektaş S, Kiliçarslan A. Papular elastorrhexis. Cutis 2013;92:E4-5.
5. Flores PB, Cuevas J, Sánchez C, De Eusebio E, Vergara A. Papular elastorrhexis: An acquired disorder of elastic tissue. Eur J Dermatol 2010;20:525-6.
6. Zhao JQ, Xu Z, Wang B, Li YJ, Zhang GQ. Dermoscopy of papular elastorrhexis. Chin Med J (Engl) 2021;134:1503-4.
7. Lewis KG, Bercovitch L, Dill SW, Robinson-Bostom L. Acquired disorders of elastic tissue: Part II. Decreased elastic tissue. J Am Acad Dermatol 2004;51;165-85.
8. Choi Y, Jin SY, Lee JH, Kwon HB, Lee AY, Lee SH. Papular elastorrhexis: A case and differential diagnosis. Ann Dermatol 2011;S53-6. doi: 10.5021/ad.2011.23.S1.S53.
9. Xue R, Yuan L, Huang H. Papular elastorrhexis. Indian J Dermatol Venereol Leprol 2015;81:529-31.

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