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CASE REPORT

PAPULAR ELASTORRHEXIS: AN UNWONTED BENIGN DISORDER OF ELASTIC TISSUE

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ABSTRACT:

Papularelastorrhexis (PE) is a rare acquired disorder of elastic tissue, which is characterized by multiple, non-follicular, skin coloured to white papules that predominantly affects trunk and upper extremities. Till date, only 31 cases have been reported making PE an extremely rare entity. The lesions are usually not symptomatic and gradually evolve over years. PE has no established treatment guidelines and most cases do not cause symptoms significant enough to warrant treatment.

Keywords :popular elastorrhexis, elastic tissue, rare disease, connective tissue

INTRODUCTION

Papularelastorrhexis (PE) is anunwonted acquired disorder of elastic tissue of the integumentary system, which is typified by variable number ofnon-follicular skin coloured to whitish papules that are predominantly located over upper trunk and upper limbs. The papules appear in adolescence without any prior inflammation or trauma over a normal looking skin.(1)The foremost report of papularelastorrhexis was given by

Bordas *et al.* in 1987.(2)Hitherto, only 31 cases have been reported all over the world, of which 11 were males and 20 were females, thus showing a female predominance.(3) To the best of our knowledge, based on expansive search of the existing databases, only one such case of PE has been reported from India in the past one decade, thus making PE an extremely rare entity.(1)Possibly, the incidence of PE is underreported and underestimated as the condition goes undiagnosed sometimes since the lesions are usually subtle, asymptomatic, benign and might be confused with other dermatoses. Treatment of PE is a matter of debate and no curative option exists till now. (4)

CASE HISTORY

A 23-year-old differently abled gentleman, reported to the author(s) with complaints of scattered, flesh coloured asymptomatic papular eruptions over the left side of the neck, since the past one year.**[FIGURE 1]** The lesions which were several millimeters in size, were first noticed by his sister who brought him to author(s) and was the informant. They were insidious in onset and they progressively increased in size and number. There was no history of trauma or any history suggestive of any inflammatory process in the region. There was no history of similar illness in the family.Cutaneous examination revealed multiple, 1-3 mm sized, well defined, soft,compressible, skin coloured, non-follicular papules over the left side of the neck. General physical examination revealed that he is moderately built and nourished with stable vitals. No other significant lesions were seen on his nails, hairs, mucosa or any other part of the skin. Routine blood tests were within normal limits. Although we advised biopsy on the first visit, his sister was reluctant. Therefore, we advised him to follow up after one month and asked him to apply a bland emollient. The patient reported duly after one month. The lesions had increased in number and his family consented for biopsy. A 4mm punch biopsy of one of thelesions was taken from the left side of his neck. Histological examination with hematoxylin and eosin staining revealed foci of altered collagen in upper dermis without any inflammatory infiltrate.**[FIGURE 2]** Special stainfor detecting elastic tissue (GomoriAldehyde-Fuchsin stain) was done which showed near absence of elastic tissue within such areas. Thus, a final diagnosis of PE was made. The family was counseled and reassured that no

treatment was required for the lesions in view of their asymptomatic nature and was advised follow up.

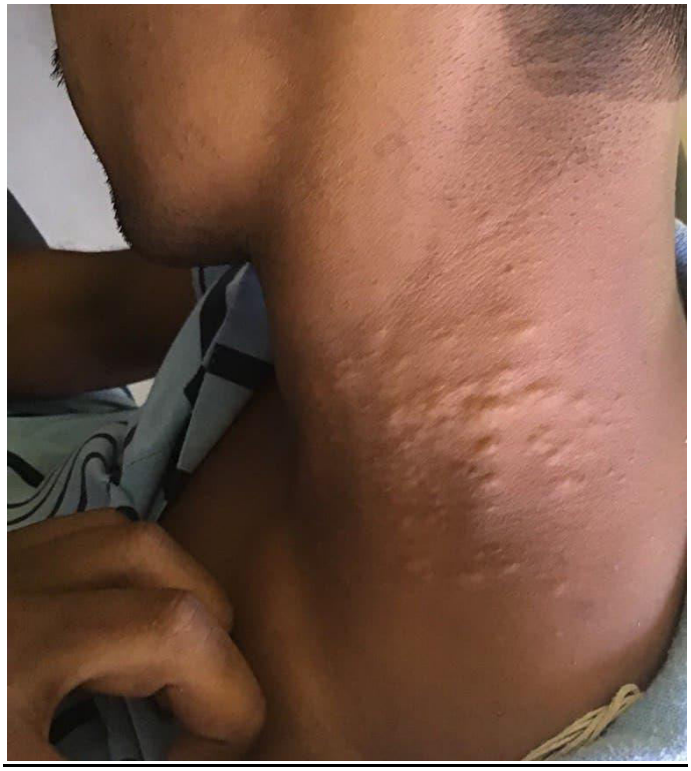

Figure 1: Flesh-coloured asymptomatic papular eruptions over the left side of the neck.

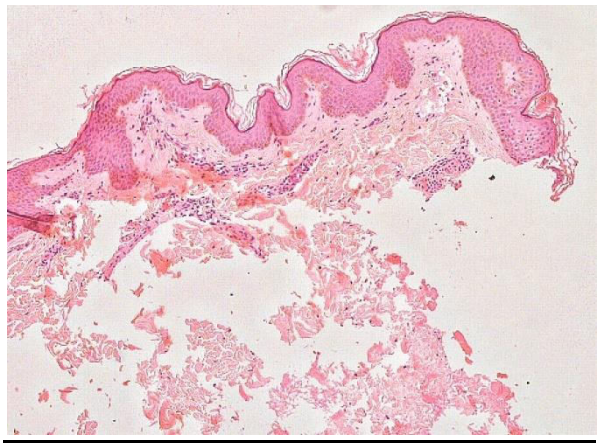

Figure 2: Histological examination with hematoxylin and eosin staining of tissue from the affected area showing foci of altered collagen in the upper dermis without any inflammatory infiltrate.

DISCUSSION

PE is a rare cutaneous disorder which causes aberration in elastic tissue.(2) No associated extracutaneous manifestations are usually seen. No inheritance pattern has been reported yet. The etiopathogenesis is not fully known. All the reported cases weresporadic and acquired. Some authors consider it as an incomplete variant of Buschke-Ollendorff syndrome, while some say that it is related to immunological imbalances causing disparity between upregulated T-cell mediated catabolic pathways and elastic anabolic mechanisms.(5)The pivotal histological feature is decrease or fragmentation of dermal elastic fibres with or without changes in collagen bundles in the dermis.(6)(7)Clinical and histopathological differential diagnoses of PE are many and includes nevus anelasticus, eruptive collagenoma,anetoderma, mid dermal elastolysis, cutis laxa, papular acne scars, disseminated lenticular dermatofibrosis (as a component of Buschke–Ollendorff syndrome), white fibrous papulosis of the neck, pseudoxanthoma elasticum, pseudoxanthoma elasticum-like papillary dermal elastolysis, mid-dermal elastolysis, and perifollicular elastolysis.It is still a matter of controversy whether PE is a separate entity or it belongs to nevus anelasticus or Buschke-Ollendorf syndrome. Bordas *et al.* suggested that PE was a variant of nevus anelasticus due to the reduction and fragmentation of elastic fibers, while Schirren *et al.*described a family with PE and proposed that it was an abortive form of Buschke-Ollendorf syndrome. (8) Ryder *et al.* supported the theory that eruptive collagenoma, nevus anelasticus and PE represents one disease or disease spectrum.(9) A final diagnosis of PE is important since a long list of differential diagnosis exist and once the diagnosis is confirmed only reassurance and counselling of the patient is required.There are reports demonstrating that intralesional injections of triamcinolone acetonide causes improvement of the lesions. (10) But PE has no established treatment guidelines and most cases do not cause symptoms significant enough to warrant treatment.

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