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REVIEW

Papular elastorrhexis: clinical perspectives

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Abstract: First described by Bordas in 1987, papular elastorrhexis (PE) is a rare elastic fiber disorder of the skin characterized by multiple, discrete, asymptomatic, firm, nonfollicular, monomorphous, 1–5 mm, circumscribed, hypopigmented, oval to round papules, symmetrically distributed on the chest, abdomen, back, shoulders, arms, and thighs. The onset of the condition is usually in the first or second decade of life. PE appears to be an exceedingly rare entity, with 33 cases reported in the literature until now. However, the disorder might be underestimated probably because of its subtlety, asymptomatic course, and benign nature of clinical alterations, which can easily be confused with other dermatoses such as acne scars. Clinical and histopathological differential diagnosis of PE is broad and includes papular acne scars, eruptive collagenoma, disseminated lenticular dermatofibrosis (as a component of Buschke–Ollendorff syndrome), white fibrous papulosis of the neck, pseudoxanthoma elasticum, pseudoxanthoma elasticum-like papillary dermal elastolysis, middermal elastolysis, and perifollicular elastolysis. Treatment of PE is a matter of debate and no reliable curative option exists.

Keywords: papular elastorrhexis, elastic fibers, connective tissue

Introduction

Papular elastorrhexis (PE) is a rare disorder clinically characterized by asymptomatic papules that show fragmentation of dermal elastic fibers on biopsy.[1] The lesions present as multiple, nonfollicular, hypopigmented papules, 1–5 mm in size located mainly on the chest, back, and abdomen. The onset is usually within the first or second decade of life. Histopathological examination reveals focal fragmentation and decrease of elastic fibers in the dermis. Treatment of this entity is a matter of debate since no effective treatment option exists in the literature. However, definitive diagnosis of PE and reassurance of the patients regarding the benign nature of this entity are important. In this manuscript, epidemiological, clinical, histopathological portrait and differential diagnosis of PE is discussed using a PubMed search.

History

PE was initially described in 1987 by Bordas et al[2] as a variant of nevus anelasticus. The clinical presentation in the original report was characterized by several small yellowish papules on the trunk with a reduction and fragmentation of the elastic fibers histopathologically. In 1988, Sears et al[3] presented two additional cases and considered that the entity may represent a variant of connective tissue nevus.

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Epidemiology

Up to now, 33 cases of PE have been reported in the English literature.[4–9] The rarity of this disease is probably due to its subtlety, asymptomatic course, and benign nature of clinical alterations, which can easily be confused with other dermatoses such as acne scars.[7,10] The condition has a predilection for the female gender, with 20 female and 13 male patients reported so far.[9] The onset is usually in the first or second decade of life.[1,11] However, a single case has been reported in a 45-year-old woman.[1]

Etiology and pathogenesis

The etiopathogenesis is skeptical.[1] Except for an isolated case of familial clustering, most of the cases reported hitherto are sporadic and acquired.[1,4,10] There is typically no history of antecedent trauma, local inflammation, infection, or acne.[1,5,6]

The nosological classification of PE is also controversial.[4] Some authors consider familial PE as an abortive incomplete variant of Buschke–Ollendorff syndrome (BOS) without associated osteopoikilosis.[3,12] BOS is an autosomal dominant disease characterized by disseminated connective tissue nevi (usually elastomas) or osteopoikilosis or both and is associated with mutations in the *LEMD3* gene.[1,5,16] In one study, *LEMD3* mutations have not been identified in two patients with PE, and the authors concluded that PE does not belong to the genomic spectrum of BOS.[4] Thus, some authors believe that PE is a unique entity distinct from BOS.[4–6,8] Nevertheless, PE is a very rarely encountered disorder, and the debate as to whether it is a variant of BOS or a distinct entity continues.[4]

A single case report of sudden onset of PE in a 13-year-old child during immunological recovery from HIV infection has been reported. The authors hypothesized that the sudden immunological recovery may trigger an imbalance between elastin anabolic mechanisms and T-cell upregulated catabolic pathways, which could have caused the marked loss and fragmentation of elastic tissue.[13]

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.[14]

Some authors consider PE as a reparative process based on clinical and histopathological findings.[3] Reports of PE localized to sites of antecedent trauma may support this theory.[15]

Clinics

Clinically, the disorder is characterized by multiple, discrete, asymptomatic, firm, nonfollicular, monomorphous, 1–5 mm, circumscribed, oval to round papules.[1,5,12] The lesions have no tendency to group or merge into plaques.[5,12] They are typically skin colored or creamy, although hypochromic variants have been reported.[3,7,8] The lesions are symmetrically distributed on the chest, abdomen, back, shoulders, arms, and thighs.[1,5] There have been anecdotal reports demonstrating uncommon locations such as the armpits, neck, nape, occipitocervical, and mandibular regions.[5,7] A solitary variant has been reported.[9] The lesion was described as a single, well-defined, soft, hypopigmented, nonfollicular papule of 0.5 cm diameter with a wrinkled surface in a 16-year-old boy with diagnostic histopathological features. An unusual case of several flesh-colored papules on the wrist of a 54-year-old woman, corresponding to an intravenous infusion drug site, has also been described.[15] Facial PE and PE developing in an eruptive manner have been described by our group.[1,5,16] So far, there has been no report of systemic affection.[1,5,12]

Histopathology

Histopathological examination displays substantial focal fragmentation and decrease or partial loss of elastic fibers in the reticular dermis, which results in a speckled appearance.[1,10] A slight perivascular lymphohistiocytic superficial and deep dermal inflammation might be observed.[1,10] There might be focal areas of fibrosis in the middermis, where collagen bundles may be thickened, homogenized, and condensed.[1,4,5] Mucin deposition around the abnormal elastic fibers has rarely been documented.[17]

Electron microscopy may reveal an absolute reduction of elastic tissue, with a relative increase in the fibrillar component of elastic fibers as compared with normal fibers.[1,7]

Prognosis

After a gradual evolution phase over the years, the lesions are relatively stable, with no tendency for spontaneous resolution.[7]

Differential diagnosis

The list of differential diagnostic considerations embraces nevus anelasticus, papular acne scars, eruptive collagenoma, disseminated lenticular dermatofibrosis (BOS), white fibrous papulosis of the neck, pseudoxanthoma elasticum, pseudoxanthoma elasticum-like papillary dermal elastolysis, middermal elastolysis, and perifollicular elastolysis.[1,6–8]

Table 1 summarizes the key epidemiological, clinical, and histopathological differential diagnosis of PE.

Table 1 Differential diagnosis of papular elastorrhexis

Entity	Age	Clinics	Histopathology	Associations
Papular elastorrhexis	First to second decades	Multiple, discrete, nonfollicular, hypopigmented, 1–5 mm papules on the chest, abdomen, and back	Focal fragmentation decrease or loss of elastic fibers Thickened collagen bundles Perivascular inflammation	None
Nevus anelasticus	Few cases reported	Perifollicular papules with a pink to red hue Confluent plaques	Fragmentation (less prominent) or loss (more prominent) of elastic fibers Minimal or no changes in collagen bundles	None
Acne scars	Second decade	Asymptomatic, hypopigmented papules on the upper back, shoulder, and chest	Scarring with fibroblastic proliferation and elastic tissue alteration around pilosebaceous unit	Acne
Eruptive collagenoma	First to second decades	Multiple, white or flesh-colored, 2–5 mm papules on trunk	Prominent thickened, swollen, and homogenized collagen bundles with reduced elastic fibers	Down syndrome MEN type 1
Dermatofibrosis lenticularis (BOS)	Childhood	Asymmetrical, skin-colored papules and plaques on the trunk and extremities	Thickened and branched elastic fibers encircle the collagen bundles	Osteopoikilosis
White fibrous papulosis of the neck	Fourth to eighth decades	Symmetrical, 2–3 mm, white to pale papules restricted to neck and upper sternal region	Marked fibrosis and altered elastic tissue in the upper dermis	None
Pseudoxanthoma elasticum	First to second decades	Small, yellowish papules on the neck, axilla, and groins	Basophilic thickened elastic fibers identified on H&E stains	Eye and cardiovascular involvement
Pseudoxanthoma elasticum-like papillary dermal elastolysis	Sixth to eighth decades	Asymptomatic or pruritic, yellow or skin-colored, 2–3 mm papules on the lateral and posterior aspects of the neck, axilla, and arms	Bandlike loss of elastic fibers restricted to papillary dermis	None
Middermal elastolysis	Third to fifth decades	Plaques revealing fine wrinkling on the trunk, neck, and arm	Loss of elastic tissue restricted to middermis	Urticaria Granuloma annulare
Perifollicular elastolysis	A few cases reported	Gray to white papules on the head, upper trunk, and arms	Elastic fiber alteration centered around hair follicles	Behçet's disease Atopic dermatitis

Abbreviations: BOS, Buschke–Ollendorff syndrome; MEN, multiple endocrine neoplasia.

Treatment

Affected patients should be reassured about the benign nature of the condition.[7] There is no reliable curative treatment for papular elastorrhexis.[1,5] One anecdotal report showed improvement after intralesional injection of triamcinolone.[1] Oral antibiotics, oral isotretinoin, topical tretinoin, and benzoyl peroxide usually have futile outcomes.[1,3,11]

Conclusion

PE is a rare elastic fiber disorder of the skin with a predilection for the female gender, and its onset is in the first or second decade of life. The disorder is characterized by multiple, discrete, asymptomatic, nonfollicular, monomorphous, hypopigmented, oval to round papules symmetrically distributed on the chest, abdomen, back, shoulders, arms, and thighs. The ultimate diagnosis of this rare entity is important, since a broad list of differential diagnostic considerations exists. Thorough knowledge of its epidemiological, clinical, and histopathological features and exclusion of associated systemic disorders are essential to reach the final diagnosis. Then, reassurance of the patients will be all that is required.

The limitation of this study is the absence of meta-analysis, because of the rarity of the condition and lack of clinical cohort studies to obtain a meta-analysis. Thus, the information presented herein is based on case presentations of the disorder, derived from a complete PubMed search.

Disclosure

The authors report no conflicts of interest in this work.

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