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CASE REPORT

Multiple arcuate lesions over neck of an adult: a case of atypical Elastosis Perforans Serpiginosa

Múltiplas lesões em arco no pescoço de um adulto: um caso atípico de Elastose Perfurante Serpiginosa

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Abstract

Elastosis perforans serpiginosa (EPS) is a rare disorder of the skin, usually present in early adulthood over the neck, upper arms, or face, associated with various genetic, cardiac, and renal comorbidities. A 38-year-old male presented with multiple painless, intensely itchy lesions over the neck for 6 months with insidious onset and gradually progressive number. There was no significant drug history or family history, or associated comorbidities or history of any intellectual disability. On examination, multiple non-tender arcuate plaques were present over the nape of the neck, without any scaling or ulceration. Due to a diagnostic dilemma, a biopsy was done, which showed elongated rete ridges forming a channel containing basophilic debris with numerous thick twisted wavy fibers with lymphocytic infiltrates. Verhoeff-Van Gieson staining came positive for elastic fibers. Hence, the case was diagnosed as idiopathic elastosis perforance serpiginosa. Lesions resolved after application of 0.05% tretinoin cream for 3 months. EPS is an uncommon disease of childhood or early adulthood, usually asymptomatic and associated with various genetic, cardiac, or renal comorbidities or drug intake. But here, lesions appeared at late adulthood, were intensely pruritic with no associated factors. Hence, the case was atypical in terms of onset, symptoms, and causal association; hence presented here.

Keywords: Elastosis perforans serpiginosa. Arcuate. Elastin. Verhoeff-Van Gieson. Basophilic debris.

Resumen

A elastose perfurante serpiginosa é uma doença rara da pele que geralmente se manifesta no início da idade adulta, afetando o pescoço, os braços superiores ou o rosto, e está associada a várias comorbidades genéticas, cardíacas e renais. Um homem de 38 anos apresentou múltiplas lesões indolores e intensamente pruriginosas no pescoço há seis meses, de início insidioso e número progressivamente crescente. Não havia histórico significativo de uso de medicamentos, antecedentes familiares, comorbidades associadas ou deficiência intelectual. Ao exame físico, observaram-se múltiplas placas arqueadas não dolorosas na nuca, sem descamação ou ulceração. Devido ao dilema diagnóstico, foi realizada biópsia que revelou cristas epidérmicas alongadas formando um canal contendo detritos basofílicos, com numerosas fibras espessas, torcidas e onduladas, além de infiltrado linfocitário. A coloração de Verhoeff-Van Gieson foi positiva para fibras elásticas. Assim, o caso foi diagnosticado como elastose perfurante serpiginosa idiopática. As lesões regrediram após aplicação de creme de tretinoína a 0,05% por três meses. A elastose perfurante serpiginosa é uma doença incomum da infância ou início da idade adulta,

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geralmente assintomática e associada a diversas comorbidades genéticas, cardíacas, renais ou ao uso de medicamentos. No entanto, neste caso, as lesões surgiram tardiamente na idade adulta, eram intensamente pruriginosas e sem fatores associados, configurando, portanto, uma apresentação atípica quanto ao início, sintomas e causa.

Palavras-chave: Elastose perfurante serpiginosa. Arqueado. Elastina. Verhoeff-Van Gieson. Detritos basofílicos.

Introduction

Elastosis perforans serpiginosa (EPS) is a rare perforating dermatosis characterized by transepidermal elimination of altered elastic fibers from the dermis[1]. It belongs to the primary perforating disorders, where degenerated elastic tissue is extruded through epidermal channels, forming distinctive lesions[2]. EPS typically presents in childhood or early adulthood with a male predominance and an estimated prevalence of < 1 in 100,000[3]. Clinically, it manifests as asymptomatic, hyperkeratotic papules arranged in arcuate, serpiginous, or annular patterns, commonly on the neck, face, or upper extremities[4]. Lesions are often self-limiting but may persist or recur.

EPS is classified into idiopathic (50-70% of cases), reactive (associated with connective tissue disorders such as pseudoxanthoma elasticum, Ehlers–Danlos syndrome, or Down syndrome), and drug-induced (commonly linked to long-term penicillamine therapy)[5-7].

Associated comorbidities include cardiovascular anomalies, renal diseases, or intellectual disabilities in syndromic cases[8]. Histopathologically, EPS shows acanthosis, hyperkeratosis, and transepidermal channels containing basophilic debris and fragmented elastic fibers, confirmed by Verhoeff-van Gieson (VVG) staining[9]. The etiology involves altered elastin metabolism, possibly due to genetic mutations or external triggers affecting dermal fibroblasts[3]. Diagnosis is clinicopathologic, as clinical mimics include granuloma annulare, porokeratosis, or reactive perforating collagenosis[10]. Treatment is challenging, with topical retinoids, cryotherapy, or laser ablation used anecdotally[11,12]. This case report describes an atypical presentation of idiopathic EPS in a middle-aged adult, highlighting diagnostic challenges and therapeutic success.

Case report

A 38-year-old male presented to the dermatology outpatient department with multiple solid lesions on the neck for 6 months. The lesions were insidious in onset, gradually increasing in number, painless but intensely pruritic, impacting daily activities. He denied preceding trauma, drug intake (including penicillamine), family history of similar lesions, or systemic symptoms such as joint pains or cardiovascular issues. No comorbidities, including connective tissue disorders or genetic syndromes, were reported. Examination revealed multiple non-tender, arcuate plaques bilaterally over the nape of the neck, surrounded by 2-3 mm keratotic papules in a serpiginous pattern, without scaling, ulceration, or erythema (Fig. 1A and B). No lesions were noted elsewhere. Differential diagnoses included granuloma annulare, annular elastolytic giant cell granuloma, and porokeratosis, but the morphology suggested a perforating disorder.

Laboratory investigations, including complete blood count and renal function tests, were normal. A punch biopsy from a lesion showed irregular acanthosis with elongated rete ridges forming narrow channels containing basophilic debris and thick, twisted elastic fibers (Fig. 1C). The dermis had mild lymphocytic and plasma cell infiltrates (Fig. 1D). Hematoxylin and eosin staining confirmed these findings, and VVG staining highlighted fragmented elastic fibers within the channels, consistent with EPS[9]. No collagen perforation or foreign material was seen. Idiopathic EPS was diagnosed based on clinical and histopathological correlation.

Topical 0.05% tretinoin cream was applied nightly. Pruritus improved within 2 weeks, and lesions resolved completely after 3 months, with no scarring or recurrence at 6-month follow-up.

Discussion

This case is atypical due to its late onset, intense pruritus, and idiopathic nature without associated factors. EPS typically presents before the age of 20 years, with only 15% of cases occurring after 30, often in reactive or drug-induced forms[3]. A systematic review of 68 EPS cases noted late-onset cases are rare and usually linked to comorbidities[3]. The intense pruritus here contrasts with the typically asymptomatic or mildly itchy EPS; pruritus is more common in acquired perforating dermatoses such as those associated with

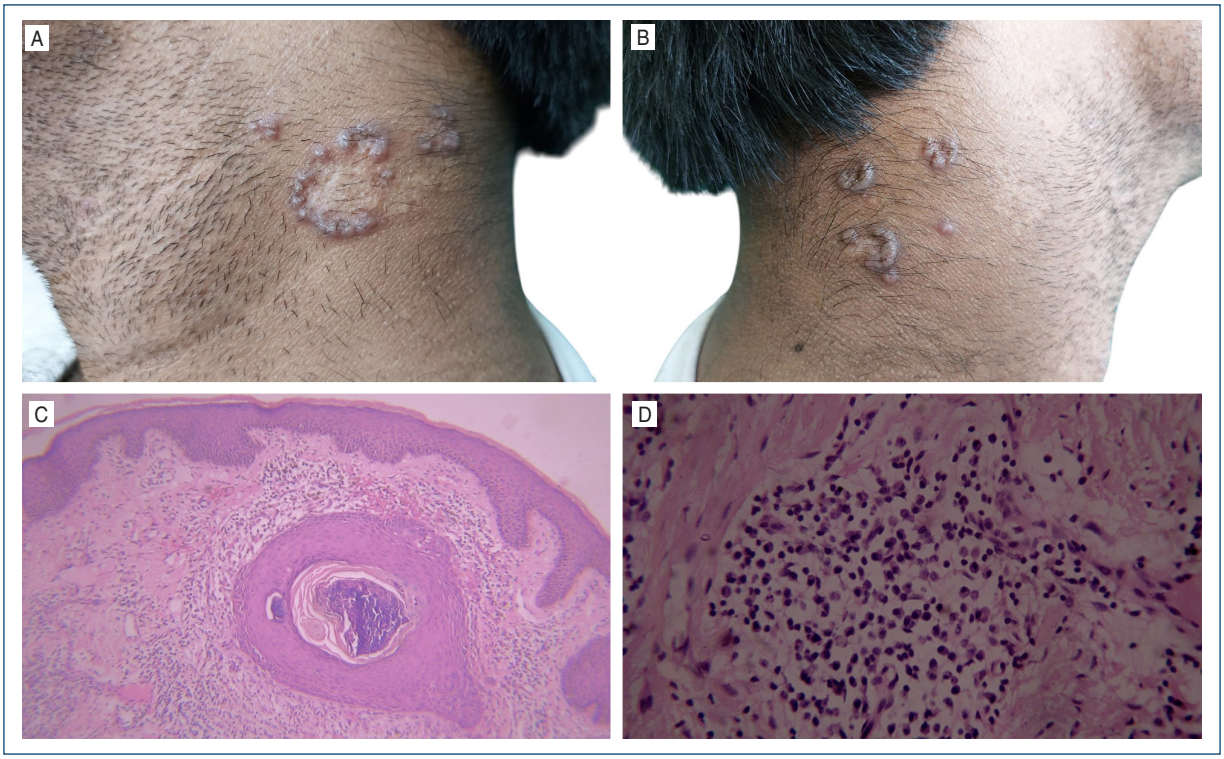

Figure 1. A and **B:** multiple arcuate plaques bilaterally over the nape of the neck, surrounded by 2-3 mm keratotic papules in a serpiginous pattern. **C:** ×10 magnification showing acanthosis, hyperkeratosis and transepidermal channels containing basophilic debris and fragmented elastic fibers. **D:** ×40 magnification showing mild lymphocytic and plasma cell infiltrates in dermis.

diabetes or renal failure, absent in this patient[2,4]. The dermal lymphocytic and plasma cell infiltrates suggest a heightened inflammatory response, though not typical for EPS[9].

The absence of genetic, drug-related, or systemic associations marks this as a rare idiopathic variant. Approximately 25-40% of EPS cases relate to genetic disorders such as Down syndrome, where elastin synthesis may be disrupted[8,13]. Drug-induced EPS, comprising 20-30% of cases, is strongly associated with penicillamine, which alters elastin cross-linking[5,6,14]. Reactive EPS may occur with pseudoxanthoma elasticum or scabies, triggering elastic fiber degradation[7,10]. This patient's lack of such factors suggests a sporadic elastin gene mutation or unidentified environmental trigger[5].

Histopathology was crucial, showing classic EPS features: transepidermal channels with elastic debris, confirmed by VVG staining[9]. This distinguishes EPS from reactive perforating collagenosis, which involves collagen[2]. Topical tretinoin led to complete resolution, consistent with its role in promoting epidermal turnover and elastic fiber clearance[11,12]. Alternatives such as cryotherapy or CO_2 laser are less effective, and systemic isotretinoin is reserved for refractory cases[4,11]. In drug-induced EPS, discontinuing the offending agent is critical, though lesions may persist[6]. Rare paraneoplastic associations with malignancies like multiple myeloma necessitate systemic evaluation in atypical adult-onset cases[7].

This case underscores the diagnostic challenge of atypical EPS, where biopsy is essential. Future research should explore genetic profiling to clarify idiopathic cases and optimize therapies.

Conclusion

This late-onset, pruritic, idiopathic EPS case without comorbidities highlights the variability of this rare dermatosis. Histopathologic confirmation and topical tretinoin therapy led to resolution, emphasizing conservative management. Clinicians should consider EPS in adults with serpiginous neck lesions, even without classical associations, to avoid misdiagnosis. This report adds to the literature on atypical EPS, advocating for increased awareness in dermatologic practice.

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Conflicts of interest

None.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the ethics committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

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