Growing Periumbilical Plaque: A Case of Perforating Calcific Elastosis

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PRACTICE **POINTS**

- Perforating calcific elastosis (PCE) is a rare, localized, acquired variant of the inherited connective tissue disorder pseudoxanthoma elasticum (PXE).
- Histopathologic findings are identical for PCE and PXE, warranting differentiation via thorough clinical examination as well as further investigation of the patient's medical and family history.
- Although there are no definitive treatments, most cases of PCE resolve spontaneously.
- Dermatologists should be aware of the importance of clinically differentiating PCE from PXE to prevent extensive workup, which can lead to unnecessary testing and increased morbidity in patients.

To the Editor:

Pseudoxanthoma elasticum (PXE) is a genetic perforating dermatosis characterized by fragmentation and calcification of elastic fibers that most commonly manifests on the skin, eyes, gastrointestinal tract, or cardiovascular system.[1] Classic skin findings include multiple symmetric yellowish papules favoring the flexural surfaces of the body and neck as well as the periumbilical and inguinal regions.[1,2] Many life-threatening complications from this disease can occur due to calcification of elastic fibers in other parts of the body, such as the internal elastic lamina of arteries, which can cause gastrointestinal tract bleeding and accelerated cardiovascular disease including valvular disease.[2,3] If PXE is localized to the skin only without systemic involvement or a family history, a diagnosis of perforating calcific elastosis (PCE) can be made. We report a case of PCE in a patient with a growing umbilical lesion.

A 49-year-old multiparous (gravida 3, para 3) woman presented for evaluation of an evolving periumbilical lesion of 4 months' duration. She denied pain, bleeding, or drainage from the area, as well as any systemic symptoms. The patient had a surgical history of a laparoscopic hysterectomy 7 years prior to the current presentation due to uterine fibroids, which resulted in a periumbilical scar. At the current presentation, physical examination revealed 2 hyperpigmented to violaceous periumbilical papules coalescing into a plaque with overlying hyperkeratosis and crusting (Figure 1). A punch biopsy was performed and histopathology showed diffuse dermal collections of degenerated eosinophilic distorted elastic fibers with calcification (Figure 2). Further sections showed a transepidermal channel in which the elastic fibers extruded from the dermis through the epidermis (Figure 3). The diagnosis of acquired PCE was made based on the clinical presentation, relevant medical history, and lack of underlying medical conditions or family history of PXE. No further workup was needed, and the patient reported no further progression and rather some improvement (decrease in size) of the lesion at 3-month follow-up.

Perforating calcific elastosis (also known as periumbilical perforating PXE) is a rare acquired condition that is seen predominantly in multiparous middle-aged women.[4-6] This diagnosis consists of degenerated calcified elastic fibers that may perforate the skin of the abdominal or periumbilical region. It clinically manifests as multiple painless hyperkeratotic papules surrounding the periumbilical region.[4-6]

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The authors report no conflict of interest.

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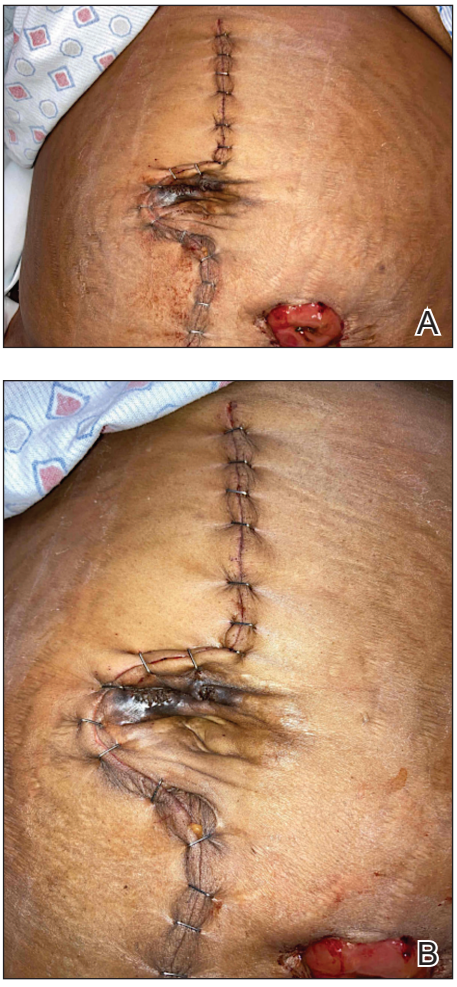

FIGURE 1. A growing hyperpigmented to violaceous periumbilical plaque with a central hyperkeratotic core that was diagnosed as perforating calcific elastosis in a patient with a history of abdominal surgery.

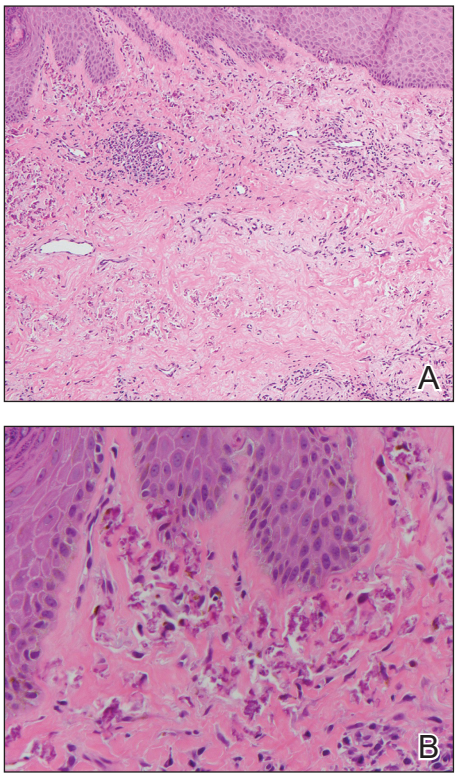

FIGURE 2. Histopathology showed diffuse dermal collections of degenerated eosinophilic distorted elastic fibers with calcification (H&E, original magnifications ×100 and ×400).

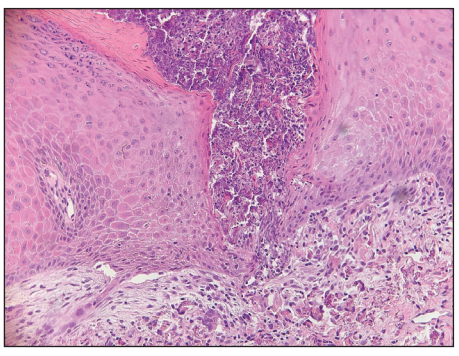

FIGURE 3. Histopathology showed a transepidermal channel extruding the dermal, eosinophilic, fragmented, curly elastic fibers through the epidermis (H&E, original magnification ×200).

The etiology and pathogenesis of PCE have not been defined but have been attributed to recurrent stressing of elastic fibers due to repeat traumas,[1] which is proposed to lead to degeneration of elastic fibers and calcification of damaged tissue.[4-7] As a result, PCE most commonly manifests in multiparous, obese, middle-aged women and patients with multiple abdominal surgeries or ascites.[1] It also has been reported in patients with renal failure due to deposition of abnormal calcium phosphate products onto elastic fibers.[4] In our patient, the development of PCE was related to both multiparity and trauma from prior surgery.

The histopathologic findings of PCE and PXE are similar, warranting differentiation via thorough clinical examination as well as further investigation of the patient's medical and family history. Both show degenerated, fragmented, curly elastic fibers with calcium deposition throughout the dermis and a transepidermal channel extruding these elastic fibers.[7,8] The biopsies stain positive for elastic fibers and calcium deposition. Calcium staining can help to differentiate these entities from elastosis perforans serpiginosa, which lacks the presence of calcium staining.[7]

There are no definitive treatments for PCE. A single case report of a patient with PCE and renal failure showed regression with hemodialysis.[9] In a study evaluating patients with inherited PXE, notable improvement was seen in skin lesions treated with bisphosphonates, possibly suggesting that regulating serum calcium may contribute to improvement of the disease.[3] Most cases spontaneously resolve with atrophic plaques. Our patient required no additional treatment with

no further progression and reported improvement of the lesion with spontaneous decrease in size.

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