Perforating Calcific Elastosis: Revisiting a Rare Entity

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Sir,

Perforating calcific elastosis (PCE), also known as periumbilical perforating pseudoxanthoma elasticum (PPPXE), is an acquired, localized cutaneous disorder, frequently found in obese, multiparous, middle-aged women.

The histopathology reveals morphologically altered and calcified elastic fibers throughout the dermis that may be extruded through the skin by transepidermal elimination. We report here a case of this rare entity occurring in a multiparous, obese woman presenting with periumbilical keratotic papules and scars showing characteristic histlogic features of PCE. A 56-year-old obese female, gravida 9, presented with a complaint of multiple itchy, raised lesions around the umbilicus for 2 years. Her ailment began as a skin-colored soft papule near the umbilicus with subsequent development of multiple similar lesions over the periumbilical region. As new lesions developed, older lesions healed with scarring. There was no history of discharge from the lesions or involvement of other sites. She was normotensive and normoglycemic. She did not give any history of difficulty in breathing or blood in vomitus, sputum, or stool. Her surgical and drug histories were also unremarkable. There was no history of similar lesions in her family.

On examination, we found multiple keratotic papules around the umbilicus, about 1 cm × 1 cm in size [Figure 1]. The lesions were skin colored while few had a violaceous tinge. Firm keratotic plug was present inside a papule. Scarring was evident in the involved area. The rest of the mucocutaneous system was normal. We kept reactive perforating collagenosis (RPC), Kyrle’s disease, elastosis perforans serpiginosa (EPS), perforating granuloma annulare, and PCE as the differentials.

Her laboratory investigations for hemoglobin, fasting and postprandial sugar, and serum calcium and phosphate were normal. Stool for occult blood and urine for red blood cells were tested negative. Her blood pressure was also within normal range, and abdominal ultrasonography and ocular examination revealed no abnormality.

A punch biopsy from a keratotic papule sent for histology with hematoxylin and eosin staining (H and E) revealed acanthotic epidermis with pseudoepitheliomatous hyperplasia [Figure 2]. There were irregularly clumped, short, fragmented, basophilic elastic fibers in the mid-dermis. Staining with Verhoeff’s stain showed the fibers to be short, curled, and thick [Figure 3] and stained positive for calcium with von Kossa stain [Figure 4]. Although actual extrusion of the altered elastic fibers could not be demonstrated in the single-biopsy specimen, areas of perforation of the acanthotic epidermis by such fibers were visualized [Figure 5].

RPC and Kyrle’s disease were excluded by the absence of transepidermal extrusion of collagen and inflammatory debris, respectively. The presence of characteristic mineralization of elastic fibers excluded EPS. Based on the presence of periumbilical keratotic papules in an obese, middle-aged multiparous woman, without a family history and systemic features of PXE, and the characteristic histopathological features, we arrived at a diagnosis of PCE. The term PCE, also referred to as PPPXE, was used to describe periumbilical keratotic papules and plaques which on histology showed curled, fragmented, and mineralized elastic fibers as seen in PXE. The etiologic nature of PCE has been debated. Some hypothesize that it is an acquired lesion developing as a consequence of local cutaneous trauma. Neldner and Martinez–Hernandez proposed the term “localized acquired cutaneous PXE,” due to the absence of systemic manifestations and lack of familial affection of hereditary PXE. PCE has also been reported in patients with metabolic abnormalities of calcium and phosphate. Others have argued that PCE may represent a localized cutaneous expression of hereditary PXE. Hypertension in three-quarters of reported patients and angioid streaks in one-third of examined patients support this argument. Therefore, patients must be thoroughly screened for systemic manifestations of hereditary PXE before arriving at a diagnosis of PCE.

Figure 1: Periumbilical keratotic violaceous papules with areas of scarring in between
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PCE begins as asymptomatic or mildly pruritic erythematous papules, plaques, or nodules. Over the years, the lesion tends to resolve, leaving a well-demarcated, hyperpigmented, reticulate, atrophic central plaque with raised, scaly border. Discrete, horny, hyperkeratotic papules may be scattered around the periphery.

The triggering factors that have been identified for PCE are obesity, multiparity, surgery, and ascites. The occurrence of PCE in patients with renal failure suggests that abnormal calcium phosphate products may produce abnormal calcification of elastic fibers. Histopathology of PCE is characterized by degenerated elastic fibers that become infiltrated with calcium in the mid-dermis. H and E-stained sections showed altered elastic fibers throughout the dermis that are short, thick, irregularly clumped, and basophilic.[4] These calcified elastic fibers can be extruded to the skin surface through a channel lined by acanthotic epidermis. Foreign-body giant cells, histiocytes, and a few chronic inflammatory cells may be seen if perforation occurs. The histological differentials for PCE are lipodermatosclerosis, morphea profunda, erythema nodosum, granuloma annulare, lichen sclerosus, tumefactive lipedema, and basal cell carcinoma.[6] Clinical features, however, can easily exclude these histological differentials.

There is no specific treatment. Topical steroids and retinoids have been tried but without benefit.[2] One patient with renal failure showed regression of lesions with hemodialysis.[7]

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 anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.
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