

## Pruritic Periumbilical Plaque as a Presentation of a Rare Perforating Dermatitis

### Abstract

Perforating dermatoses are characterized by transepidermal elimination of altered dermal components or foreign particles. Owing to their common clinical presentation as umbilicated papules with a keratotic plug, histopathology and special staining play a very crucial role in the diagnosis. Perforating calcific elastosis, (periumbilical perforating pseudoxanthoma elasticum), an uncommon acquired localized cutaneous dermatosis, is characterized by transepidermal elimination of modified elastic fibres. It is usually seen in middle-aged obese multiparous women as well-defined periumbilical hyperpigmented atrophic plaques. We report a case of a 66-year-old female who presented with a mildly pruritic hyperpigmented periumbilical plaque of 2 years duration. Histopathology studies revealed multiple fragmented, thick, short, and curly eosinophilic fibers; along with granular basophilic material in the dermis, which stained positive for calcium and elastin, thus clinching the diagnosis of perforating calcific elastosis. There were no features of hereditary pseudoxanthoma elasticum. We report this case for its rarity.

**Keywords:** Calcified elastic fibres, multiparity, periumbilical plaque, transepidermal elimination

### Introduction

Perforating dermatoses are disorders characterized by elimination of altered dermal components or foreign particles from the dermis to outside through a transepidermal channel. Classically, perforating dermatoses are classified into four distinct entities based on the primary defect and nature of dermal material or foreign substance being eliminated: i) Reactive perforating collagenosis ii) Elastosis perforans serpiginosa (EPS) iii) Perforating folliculitis iv) Kyrle's disease.<sup>[1]</sup> Owing to their common clinical presentation as umbilicated papules with a keratotic plug, histopathology and special staining play a crucial role in the diagnosis.

Disorders such as pseudoxanthoma elasticum (PXE), granuloma annulare and cutaneous tuberculosis can also exhibit the phenomenon of transepidermal elimination.<sup>[2]</sup> Perforating calcific elastosis (PCE), an atypical form of perforating dermatoses, is an acquired elastic defect found in obese, multiparous, middle-aged women characterised by

expulsion of calcified elastic fibres through a transepidermal channel.<sup>[3]</sup>

### Case Report

A 66-year-old multiparous female, gravida 6, presented with itchy pigmented lesions over the abdomen of 2 years duration with an occasional discharge of white particles from the lesion. There was no preceding trauma and abdominal surgeries. She had no other comorbidities and her family history was unremarkable. She was treated with antifungals and topical corticosteroids with no improvement. Her BMI was 21.11 kg/m<sup>2</sup>. Cutaneous examination revealed the presence of a well-defined, hyperpigmented, atrophic periumbilical plaque with keratotic papules at the margins as well as few discrete keratotic papules over the abdomen [Figure 1]. The surrounding skin was lax with multiple striae. Differential diagnosis of chromoblastomycosis, perforating dermatoses, lupus vulgaris, and actinomycosis were considered. Her complete blood count, liver and renal function tests, blood sugars, were within the normal limits. Histopathology of the keratotic papule revealed hyperkeratosis

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### Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ\_95\_19

### Quick Response Code:



**How to cite this article:** Bathina M, Hegde SP, Shanavaz AA, Saldanha PP. Pruritic periumbilical plaque as a presentation of a rare perforating dermatosis. Indian Dermatol Online J 2020;11:68-71.

**Received:** February, 2019. **Revised:** June, 2019.

**Accepted:** July, 2019. **Published:** January, 2020.

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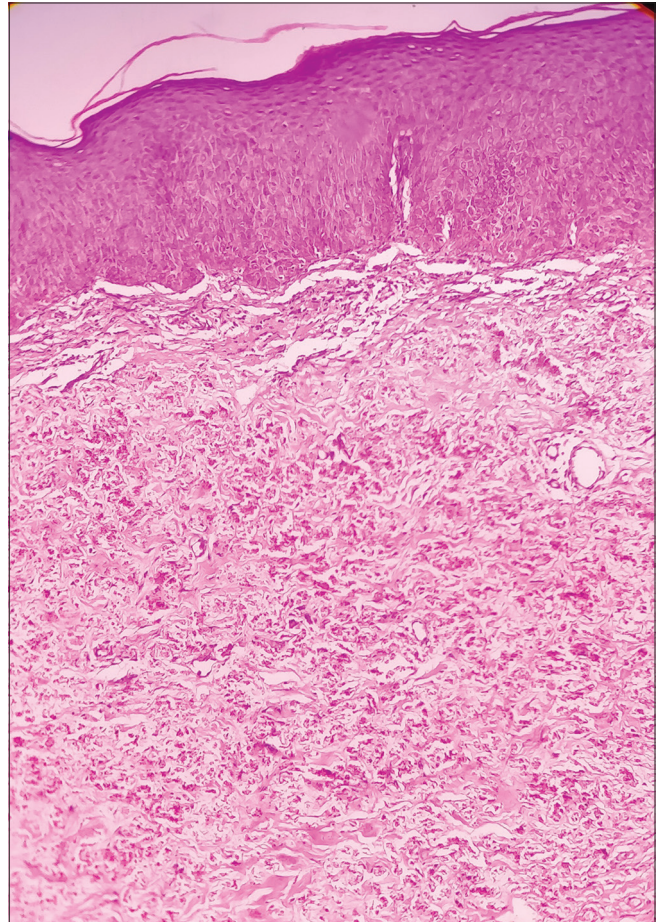
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with irregular acanthosis. Dermis showed many fragmented, thick, short, and curly eosinophilic fibers with granular basophilic material [Figure 2] and transepidermal elimination of calcified elastic fibers on haematoxylin and eosin staining [Figure 3]. The specimen also stained positive for calcium (Alizarin Red stain) [Figure 4] and elastic fibres [Verhoffs Elastic Van Gieson stain (VVG)] [Figure 5]. Bacterial and fungal cultures yielded no growth. Mantoux test was also negative. Her ophthalmological examination, chest X-ray, and abdominal ultrasonography were normal. Based on the clinical and histopathological findings, a diagnosis of perforating calcific elastosis was attained. There were no features of inherited pseudoxanthoma

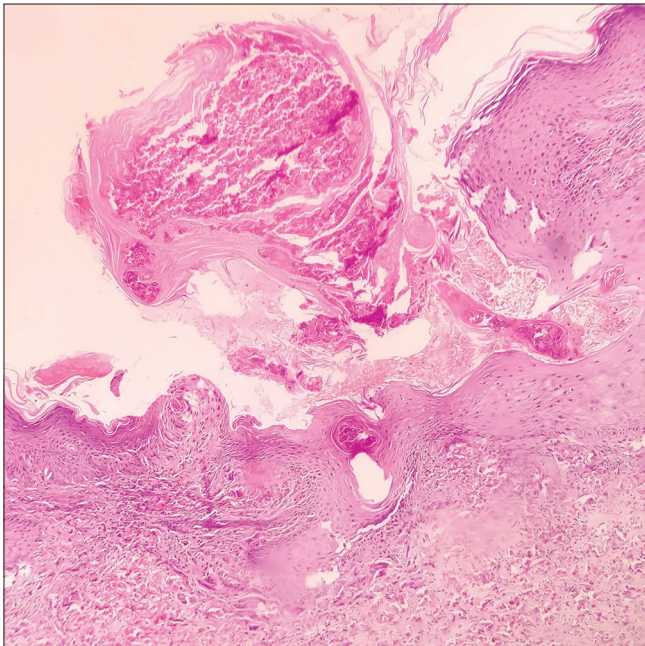
elasticum. She was treated with topical retinoids but was lost to follow-up.



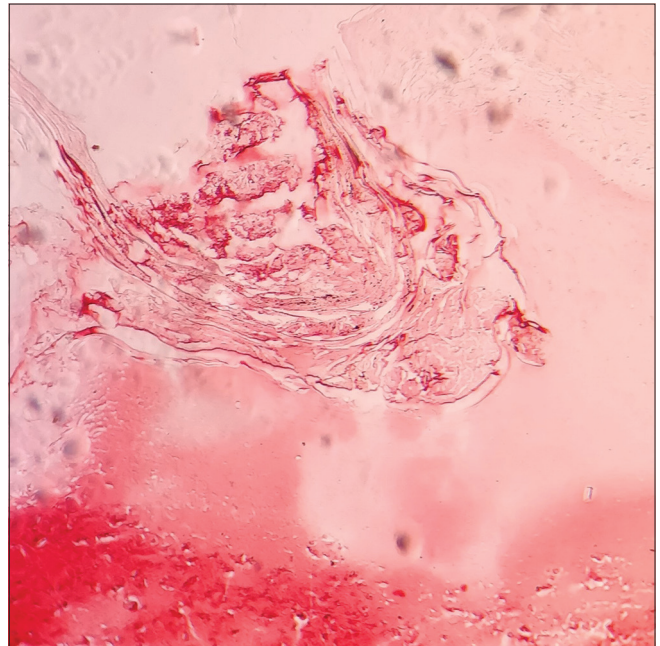
**Figure 1:** Periumbilical hyperpigmented atrophic plaque with marginal and discrete keratotic papules



**Figure 2:** Fragmented, thick, curly elastic fibres in the dermis (H and E, 10×)



**Figure 3:** Transepidermal channel with calcified elastic fibres (H and E, 10×)



**Figure 4:** Elastic fibres stained red for calcium (Alizarin Red, 10×)

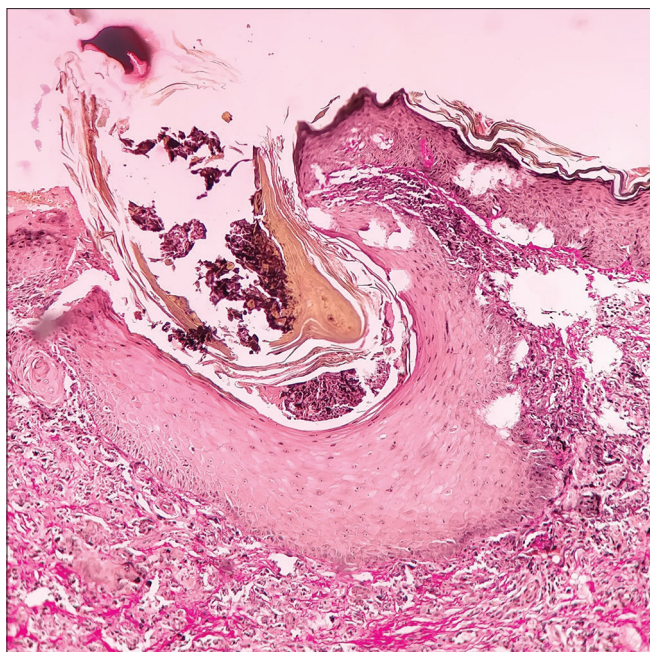


Figure 5: Elastic fibres stained black (VVG,10×)

## Discussion

Perforating calcific Elastosis, also known as periumbilical perforating pseudoxanthoma elasticum (PPPXE), was earlier thought to be associated with elastosis perforans serpiginosa (EPS). It has also been termed as “localized acquired cutaneous pseudoxanthoma elasticum” due to lack of family history and absence of systemic involvement of hereditary PXE. Some authors believe it to be a localized cutaneous feature of hereditary PXE owing to the coexistence of hypertension (75%) and angioid streaks (33%) in the reported cases.<sup>[3]</sup>

The etiology of PCE is not well established hence its pathophysiology is attributed to ectopic calcification i.e., abnormal biomineralisation of elastic fibers.<sup>[4]</sup> PCE has been most commonly reported in multiparous, obese, middle-aged women of African-American origin.<sup>[5]</sup> It may occur due to repeated cutaneous trauma, or stretching following ascites, or abdominal surgeries leading to degeneration of elastic fibres.<sup>[6]</sup> It has been observed in patients with renal failure.<sup>[3]</sup> This points toward the possibility of deposition of defective calcium phosphate products resulting in calcification of elastic tissue.<sup>[7]</sup> Application of native medications at the sites of cutaneous trauma might result in exposure to calcium salts and phosphates, and predispose to calcific degeneration of elastic fibres.<sup>[8]</sup> In our case, the cause of PCE could be attributed to the multiparous state of the patient.

Perforating calcific elastosis presents as asymptomatic or mildly pruritic gradually progressive erythematous papules or plaques in the periumbilical region or lower chest which resolve with a central well-defined reticulate

hyperpigmented atrophic plaque with scaly raised margins and peripheral discrete keratotic papules.<sup>[4,9]</sup> Clinically it can be differentiated from pseudoxanthoma elasticum by the absence of flexural and systemic involvement. The perforating disorders can thus be differentiated based on histopathology.<sup>[4]</sup>

Histopathology shows acanthotic epidermis and degenerated elastic fibres with dermal calcium deposition, mostly concentrated in the mid-dermis. Thick, short, basophilic, irregularly clumped structures i.e., elastic fibres are observed with haematoxylin and eosin staining.<sup>[4]</sup> Special stains for calcium and elastin are positive. Elastic fibres are embedded with calcium deposits. A transepidermal channel extruding this altered material is seen along with inflammatory infiltrate.<sup>[3]</sup> The histopathological differential diagnosis includes granuloma annulare, elastosis perforans serpiginosa, morphea profunda, lichen sclerosus, erythema nodosum, and lipodermatosclerosis.<sup>[7]</sup> EPS can be differentiated histopathologically from PCE by the characteristic absence of calcification of elastic fibres.<sup>[4]</sup>

This condition has no specific treatment. Topical steroids and retinoids have shown no benefit.<sup>[5]</sup>

We made a diagnosis of acquired perforating calcific elastosis based on the presence of periumbilical plaque in a multiparous woman with classical histopathological findings and the absence of features of hereditary pseudoxanthoma elasticum.

## 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 due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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