

# Periumbilical perforating pseudoxanthoma elasticum: a rare case report

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**ABSTRACT** Periumbilical perforating pseudoxanthoma elasticum (PPPXE) usually presents with well-defined periumbilical yellowish atrophic plaques with keratotic papules at the periphery. It is considered a variant of hereditary pseudoxanthoma elasticum or a localized acquired cutaneous dermatosis. The lesions usually occur in the periumbilical area in obese, multiparous women. Here, we report an additional case of periumbilical perforating pseudoxanthoma elasticum with its dermoscopic features.

## Introduction

Pseudoxanthoma elasticum (PXE) is a rare inherited disease of connective tissue causing fragmentation and mineralization of elastic fibers that primarily affects the skin, retina, and cardiovascular system [1]. Classically it is characterized by multiple asymptomatic small (1–5 mm in size), yellowish coalescing papules that are symmetrically distributed on the neck and flexural body areas, such as the axillae, antecubital fossae, periumbilical, inguinal and popliteal areas [2].

Periumbilical perforating pseudoxanthoma elasticum (PPPXE) is considered a localized variant of inherited PXE based on the presence of angioid streaks (22% of cases) and flexural lesions [3]. Some believe it to be an acquired dermatosis secondary to cutaneous trauma caused by multiple pregnancies, obesity, and multiple abdominal surgeries or trauma resulting in elastic fiber degeneration in these patients [4,5].

Sapadin et al considered PPPXE a bridge between the pure acquired form and the pure inherited form [6]. We report on a case of acquired PPPXE with dermoscopic features.

## Case Presentation

A 68-year-old multiparous woman presented with multiple yellowish, asymptomatic periumbilical lesions. The lesions had appeared two years ago and had slowly enlarged to form yellowish papules coalescing to form a plaque along with a few red-brown colored papules with central keratotic material (Figure 1).

There was no history of similar disease in the family. There was no history of any abdominal surgery. On examination, the patient was obese and had no evidence of any associated systemic disease. On cutaneous examination, multiple yellowish, asymptomatic periumbilical papules and



**Figure 1.** Periumbilical yellowish-white plaque with reddish brown keratotic papules at the periphery. [Copyright: ©2018 Jha et al.]

plaques were present with few red-brown colored papules. Dermoscopy (polarized, 10x, Dermlite DL 4; 3Gen, San Juan Capistrano, CA) revealed yellowish brown structureless areas along with semicircular, curved/serpiginous yellowish-brown lines, with few linear vessels along with a keratotic plug with central crater (Figure 2 A, B). On histopathology the yellowish papule showed an increased number of elastic fibers in the upper and mid reticular dermis. The elastic fibers were fragmented and curled giving the appearance of raveled wool. A focus of calcification was also seen (Figure 3). Histological examination was consistent with the diagnosis of PPPXE. Cardiologic evaluation (normal electrocardiogram and echocardiogram) and ophthalmoscopic examination did not show any changes. Routine laboratory tests and serum lipid profile were all within normal range.

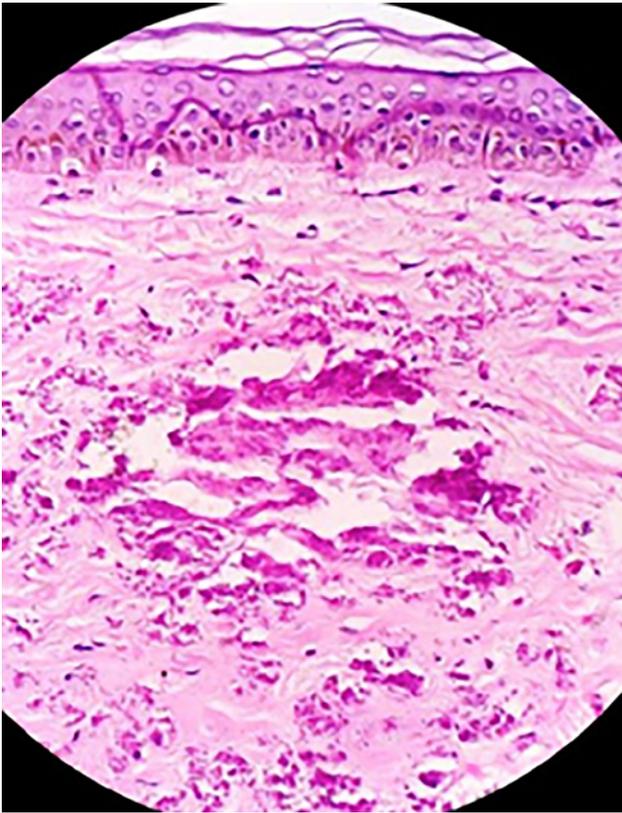
## Discussion

The term PPPXE was first used in 1979 by Hicks [4]. Earlier it was described as pseudoxanthoma elasticum (PXE) with coexisting elastosis perforans serpiginosa (EPS). Lund and Gilbert were the first to establish it as a separate entity [7]. The term “localized acquired cutaneous pseudoxanthoma elasticum” was also proposed, as PPPXE was believed to be “acquired” and lacked “systemic involvement.” [8] Recently, the term “perforating calcific elastosis (PCE)” has been sug-



**Figure 2.** (A) Dermoscopy (polarized, 10x) showed yellowish brown structureless areas along with semicircular, curved/serpiginous yellowish-brown lines, with few linear vessels. (B) Dermoscopy (polarized, 10x) showed yellowish brown structureless areas along with semicircular, curved/serpiginous yellowish-brown lines with few linear vessels and keratotic plug with central crater. [Copyright: ©2018 Jha et al.]

gested to describe this condition [9]. On dermoscopy, a yellowish hue may be due to the elastolysis of elastic fibers or the presence of calcium deposits as in PXE [10,11]. There is no established treatment of PPPXE. Dietary calcium restriction



**Figure 3.** The elastic fibers are fragmented and curled giving appearance of raveled wool with a focus of calcification. (H&E, 40x). [Copyright: ©2018 Jha et al.]

(800 mg/day) and oral phosphate binder have been reported to have significant clinical improvement in PXE [12]. These treatment modalities may be tried in PPPXE, as the pathological process involved in PXE and PPPXE is similar. We report PPPXE with calcinosis cutis in a rare presentation along with dermoscopic findings.

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