

Periumbilical and periareolar perforating pseudoxanthoma elasticum *

Pseudoxantoma elástico perfurante periumbilical e periareolar

Aline Lopes Bressan ¹

Roberto Souto da Silva ³

Alexandre Carlos Gripp ⁵

Bárbara Nader Vasconcelos ²

Maria de Fátima G. Scotelaro Alves ⁴

Abstract: The periumbilical perforating pseudoxanthoma elasticum or perforating calcific elastosis is a rare disorder and its pathogenesis is associated with the alteration of elastic fibers, which may probably result from trauma. We present the case of a 70-year-old woman with long-time blackish maculas and keratotic surface papules on the periareolar and periumbilical regions. The histopathological examination revealed altered elastic fibers, replete with calcium and formation of a path in the dermis with elastic fibers degraded in their interior, confirming the clinical diagnosis.

Keywords: Breast; Pregnancy; Pseudoxanthoma elasticum; Umbilicus

Resumo: O pseudoxantoma elástico perfurante periumbilical (PEPP), ou elastose cálcica perfurante, é distúrbio raro e sua patogênese está associada a alteração da fibra elástica, tendo o trauma provável participação. Apresenta-se caso de paciente de 70 anos com quadro de máculas enegrecidas e pápulas encimadas por crostas na região periareolar e periumbilical há mais de dez anos. O exame histopatológico revelou fibras elásticas alteradas, repletas de cálcio, e formação de pertuito na derme com fibras elásticas degradadas no seu interior. O caso descrito corresponde ao PEPP, que nesta paciente também acomete área periareolar bilateralmente.

Palavras-chave: Gravidez; Mama; Pseudoxantoma elástico; Umbigo

INTRODUCTION

The periumbilical perforating pseudoxanthoma elasticum (PPPE) is a rare acquired disease. Occurs mainly among black, multiparous, obese, and middle-aged women and in general the internal organs are not affected. Biopsy is the most important method used to make its diagnosis, and the disease has as its characteristic alteration the calcification and extrusion of elastic fibers through the epidermis.¹

We report here the case of a patient with periumbilical perforating pseudoxanthoma elasticum that, as for this patient, also strikes the periareolar region, bilaterally.

CASE REPORT

Female patient, aged 70, black and multiparous. Reports hyperchromic periumbilical and periareolar maculae that had appeared more than 10 years before and she also reports flaccidity in the abdominal wall. (Picture 1). The maculae are blackish, they have approximately 15 cm of diameter on the abdomen and 5 cm periareolar (Picture 2), bilaterally, with papules topped by yellowish crusts on loose, wrinkled skin. The patient reports light pruritus on the sites.

The histopathological exam of the papule and macula revealed depot of intumescent fibers, irregu-

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¹ On going post graduation course from the Dermatological service of Pedro Ernesto University Hospital (HUPE) – Rio de Janeiro (RJ), Brazil.

² On going post graduation course from the Dermatological service of Pedro Ernesto University Hospital (HUPE) – Rio de Janeiro (RJ), Brazil.

³ Resident of the Dermatological service of Pedro Ernesto University Hospital (HUPE) – Rio de Janeiro (RJ), Brazil.

⁴ Doctorate degree in Dermatology. Associate professor of Dermatology and responsible for the Dermatopathological sector of Pedro Ernesto University Hospital (HUPE) – Rio de Janeiro (RJ), Brazil.

⁵ Master degree in Dermatology. Assistant professor of Dermatology and responsible for the dermatologic ward of Pedro Ernesto University Hospital (HUPE) – Rio de Janeiro (RJ), Brazil.



FIGURE 1: Blackish, periumbilical and periareolar maculae, on flaccid and wrinkled skin

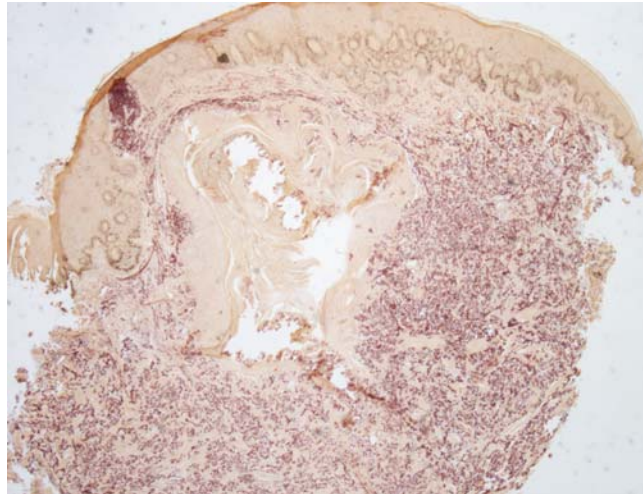


FIGURE 3: Fragmented elastic fibers evidenced by orcein coloration (100x)

larly agglomerated in the inferior middle parts of the dermis, that colored themselves as elastic fibers with orcein. (Picture 3). HE coloration showed elastic fibers slightly basophil altered as they were full of calcium and the coloration for calcium with the Von Kossa method well evidenced them (Picture 4). It was also detected an outline of a narrow passage of the dermis to the external area of the epidermis, full of degrading elastic fibers (Picture 5), that characterized the perforating form of the pseudoxanthoma elasticum or perforating calcific elastosis.

Ophthalmologic and cardiologic exams did not detect any alterations.

DISCUSSION

The periumbilical perforating pseudoxanthoma

elasticum is a rare acquired disease, with singular clinical and histological characteristics. It presents itself as a plaque restricted to the periumbilical region, of reticulated and atrophic yellowish surface, intermixed by an area of hyperchromia which gives a wrinkled aspect to the lesion. Keratotic papules are frequently found in the periphery of the lesion and can eliminate viscous material. It is more frequently found in black, multiparous, obese and middle-aged women and, in general, internal organs are not affected. Biopsy is the most important method used to make its diagnosis, and the disease has as its characteristic alteration the calcification and extrusion of elastic fibers through the epidermis.¹

In 1976, the PEPP was individualized and separated from the perforating serpiginous elastosis. It was



FIGURE 2: Blackish macula, with papules topped by crusts. (calcium extrusion)

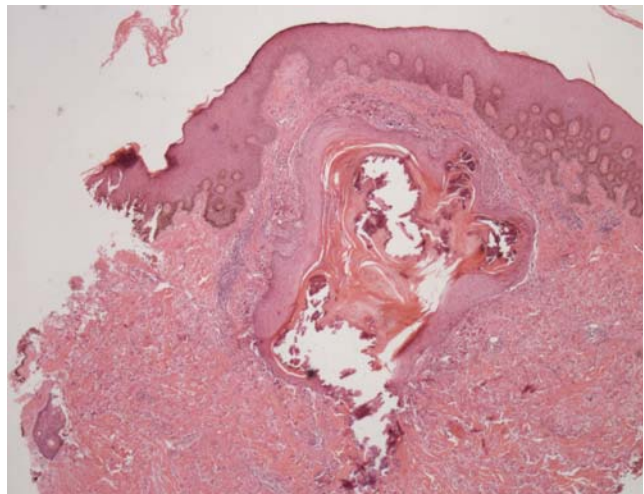


FIGURE 4: Von Kossa coloration (40x): evidencing calcium and an outline of a narrow passage to the epidermis

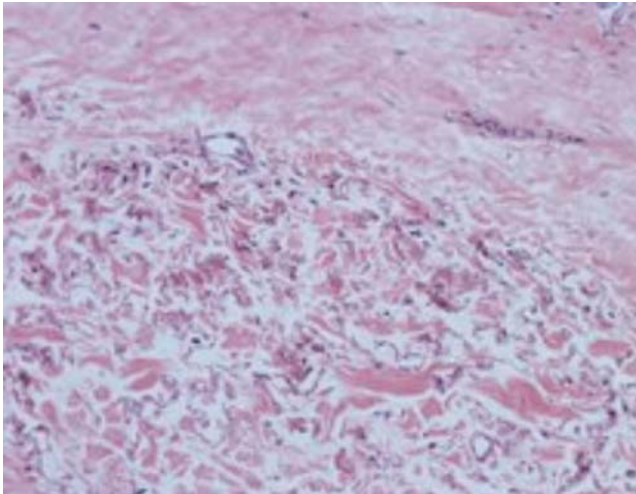


FIGURE 5: Fragmented elastic fibers. HE (200x)

proposed, in 1979, the denomination calcific elastosis to avoid the implications that the term localized PXE can offer in relation to the systemic form of the disease.²

Calcium depots are basophil and they have affinity for silver nitrate in von Kossa. coloration.

Occasionally the PXE abnormal fibers might perforate the epidermis. In some cases, the perforating PXE is only a cutaneous manifestation of the systemic hereditary form of PXE, secondarily occurring the transepidermal elimination.³ In other cases, the lesion is situated and restricted to the periumbilical region, has singular epidemiologic characteristics and occasionally might present a systemic form. In this last form, the disease has been denominated periumbilical perforating pseudoxanthoma elasticum (PPPE). The pathogenesis of PEPP is controversial. Some authors consider it acquired dermatosis resulting from cutaneous trauma by obesity, abdominal surgeries and ascites. Trauma would be an important factor in the genesis of the lesion,⁴ and its main point of alterations is on the elastic fibers.

Subsequent control includes ophthalmologic and cardiologic exams aiming at eliminating the possibility of a systemic disease.⁵ Although antiaesthetic, there is no other therapy available. If the skin redundancy is very prominent a reconstructive surgery can bring temporary relief.⁶ □

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MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:

Aline Lopes Bressan

Rua Cachambi, 34/402. Meier.

20775-182 Rio de Janeiro - RJ, Brazil

Phone: +55 (21) 2201-3590/8727-2316

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