Case for diagnosis*
*Caso para diagnóstico**

Fernanda H. M. de Souza [1] Betina Werner [2] Leila Cristina Cavalin [3]
Maurício Shigueru Sato [4] Sérgio Z. Serafini [5]

HISTORY OF THE DISEASE

Brown 74-year-old female, multiparous (10 pregnancies, 10 deliveries), with abdominal skin lesion for 16 years, on occasion pruriginous. At the physical examination, she presented two periumbilical plaques of irregular borders, yellowish color and reticular aspect, approximately 5 to 6 cm diameter, with erythematous keratotic papules (Figure 1). Absence of other lesions in the remaining tegument. Hypertension for 21 years, on a captopril and AAS regimen. Diabetes Melitus type 2, insulin-dependent for 18 years.

The histopathological exam of the supraumbilical plaque reveals reticular dermis with a large number of irregular (calcified) basophilic conjunctive fibers (Figure 2), besides an area of epidermal hyperplasia with elimination of elastic fibers through a channel filled with keratin and remnants of inflammatory cells (Figure 3).

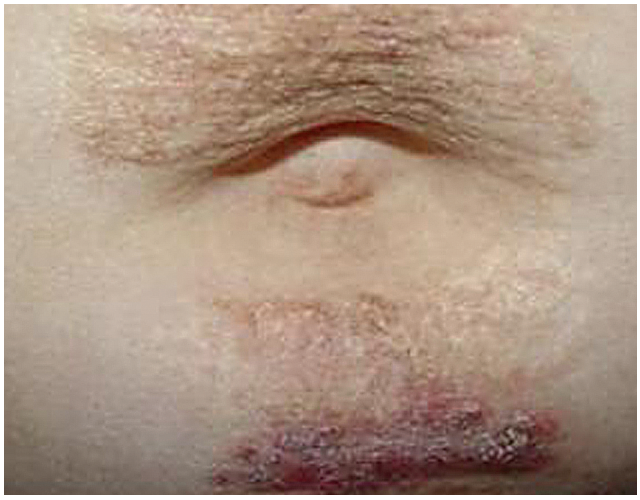

Figure 1: Two plaques with irregular borders, yellowish color, and reticular aspect around the navel, 5 to 6 cm diameter, linear erythematous keratotic papules area below the navel

COMMENTS

The Periumbilical Perforating Pseudoxanthoma Elasticum (PPPE) is an acquired defect and it is situated in the elastic tissue occurring in the periumbilical region especially of Black multiparous women around the fourth decade of life.[1,2] It is also known as Perforating Calcifying Elastosis, and it is clinically characterized by well-circumscribed or reticular yellowish plaques of irregular surface and loose consistency with diameters varying from two to 15 cm, with keratotic papules spread on their surface.[1-4] At first they are assymptomatic or slightly pruriginous and they progress for months or years. With their resolution, they leave a hyperpigmented area with a rough, inelastic irregular center. During the "perforation" phase, a necrotic material from the dermis may be extracted by means of compression of individual lesions.[1] Mechanical stress has been suggested as an important factor in the pathogenesis of this condition, as PPPE is seen predominantly in multiparous women, in patients with severe ascitis or a history of abdominal surgery, or in individuals with a genetic predisposition towards Pseudoxanthoma Elasticum.[1,2] It has also been reported in dialysis-dependent chronic renal failure patients, thus

Received on December 29, 2005.
Approved by the Consultive Council and accepted for publication on January 13, 2006.
* Work done at Serviço de Dermatologia do Hospital de Clínicas da Universidade Federal do Paraná – UFPR - Curitiba (PR), Brazil.
Conflict of interests: None

[1] 5th. Year Medical Student at the Universidade Federal do Paraná – UFPR - Curitiba (PR), Brazil.
[2] Physician, Pathologist, Doctoral student at the Hospital de Clínicas da Universidade Federal do Paraná – UFPR - Curitiba (PR), Brazil.
[3] 1st. Year Resident Doctor of the Dermatology Service, Hospital de Clínicas da Universidade Federal do Paraná – UFPR - Curitiba (PR), Brazil.
[4] Physician, 1st. Year Specialization, Dermatology Service, Hospital de Clínicas da Universidade Federal do Paraná – UFPR - Curitiba (PR), Brazil.
[5] Dermatologist, Associate Professor of Dermatology, Hospital de Clínicas da Universidade Federal do Paraná – UFPR - Curitiba (PR), Brazil.

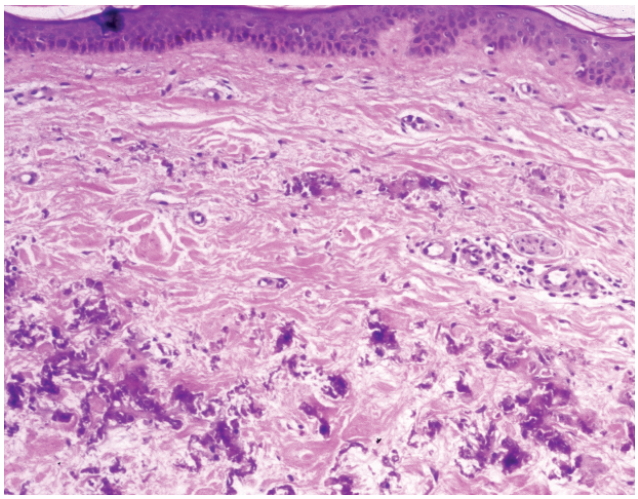

Figure 2: Irregular calcified basophilic conjunctive fibers in the reticular dermis (HE, 100x)

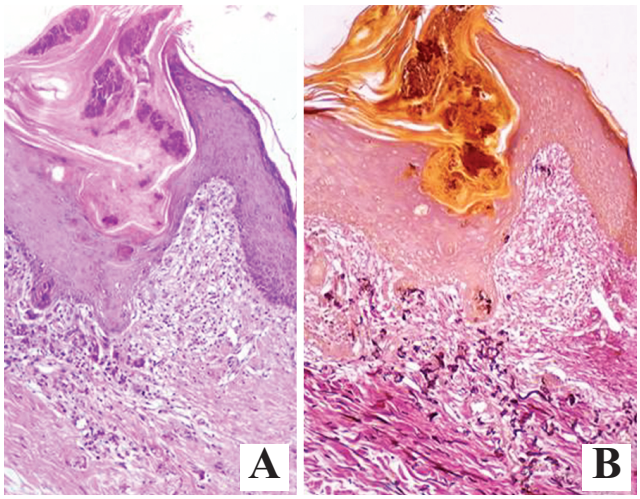

Figure 3: A: histological section of the papule area, showing epidermal hyperplasia with keratotic buffer, mononuclear inflammatory infiltrated and altered conjunctive fibers in the surface and medium dermis (HE, 100x); B: irregular elastic fibers in the dermis and epidermis in the process of elimination (Weigert, 100x)

suggesting that the abnormal metabolism of calcium and phosphate could be a predisposing factor for the development of the disease.[1,3] Histologically, PPPE is characterized by the presence of calcified, irregular basophilic elastic fibers in the reticular dermis that are eliminated through the epidermis.[2] Some have suggested its classification as a cutanous form located in the Pseudoxanthoma Elasticum, which would be acquired and would not present any systemic compromise.[1,3,4] The classic Pseudoxanthoma Elasticum is a hereditary disorder of elastic tissue with genetic heterogeneity, characterized by degeneration and calcification of elastic fibers in the ocular, cutaneous and cardiovascular systems.[3,5]

The clinical differential diagnosis should consider the annular granuloma, the chronic simple lichen, the plain lichen, sarcoidosis, and pregnancy striations. Histologically, PPPE could be indistinguishable from the classic Pseudoxanthoma Elasticum, were it not for the transepidermal elimination of abnormal elastic fibers with PPPE and the preferential situation of PE in neck and axillary folds, which thus acquire the aspect of chicken skin.

Serpiginous perforating elastosis may be differentiated by the annular, serpiginous or polycyclic configuration of the lesions, located preferentially in the neck or upper limbs, besides the fact that the altered elastic fibers eliminated through the skin are not calcified.

Besides, other "perforating" dermatoses must be considered in the histological differential diagnosis, such as the perforating annular granuloma, perforating folliculitis, and Kyrle's disease.

Several therapeutic modalities have been proposed for PPPE, such as local and systemic steroids, liquid nitrogen, tretinoin and ultraviolet light, among others,[4] and there are cases of spontaneous remission.[1] ❑

Abstract: Periumbilical perforating pseudoxanthoma elasticum is an acquired disorder of the elastic tissue found most frequently in multiparous women. A case of this condition is reported for a 74 year-old woman (G10, P10) showing yellow, reticulated plaque with erithematous and keratotic papules. Histology shows reticular dermis with irregular basophilic conjunctive fibers, as well as epidermal hyperplasia and elastic fibers elimination through keratotic channels.
Keywords: Parity; Pseudoxanthoma Elasticum; Umbilicus

***Resumo**: Pseudoxantoma elástico perfurante periumbilical é distúrbio adquirido do tecido elástico, que acomete mulheres multíparas. Apresenta-se um caso dessa condição em mulher de 74 anos (G10, P10) mostrando placa amarelada de aspecto reticular com áreas constituídas por pápulas eritêmato-ceratósicas. A histologia demonstra derme reticular com fibras conjuntivas basofílicas e irregulares, além de área de hiperplasia epidérmica, com eliminação de fibras elásticas através de canal repleto de ceratina.*
Palavras-chave: Paridade; Pseudoxantoma elástico; Umbigo

REFERENCES

1. Lewis KG, Bercovitch L, Dill SW, Robinson-Bostom L. Acquired disorders of elastic tissue: increased elastic tissue and solar elastotic syndromes. J Am Acad Dermatol. 2004; 51(Pt 1): 1-21.
2. Karp DL, O'Neil MS, Haberman AL, Taylor RM. A yellow plaque with keratotic papules on the abdomen. Arch Dermatol. 1996; 132:224-5; 227-8.
3. Sapadin AN, Lebwohl MG, Teich SA, Phelps RG, Diconstanzo D, Cohen SR. Periumbilical pseudoxanthoma elasticum associated with chronic renal failure and angioid streaks – apparent regression with hemodialysis. J Am Acad Dermatol. 1998; 39: 338-44.
4. Toporcer MB, Kantor GR. Periumbilical hyperpigmented plaque. Arch Dermatol. 1990; 126:1639-42.
5. Somasundaram V, Premalatha S, Rao MR, Razack EMA, Zahra A. Periumbilical perforating pseudoxanthoma elasticum. Int J Dermatol. 1987; 26: 536-7.

MAILING ADDRESS:
Fernanda H. Mello de Souza
Rua Gal. Polli Coelho, 355 - Tarumã
82800-180 - Curitiba – PR
Telefone: 3365-1170
Fax: 3366-4867
E-mail: nandahms@gmail.com

Dear colleague,

The "What is your Diagnosis?" section aims to present clinical cases in which the final diagnosis is questionable. If you have an article that fits this section, please contribute to the Anais Brasileiros de Dermatologia by sending it to us, our address is:

Av. Rio Branco, 39 / 18º andar - Centro - Rio de Janeiro - RJ - Brazil CEP: 20090-003