

PERIUMBILICAL PERFORATING PSEUDOXANTHOMA ELASTICUM

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A case of periumbilical perforating pseudoxanthoma elasticum occurring characteristically in an elderly, multiparous, obese, diabetic female is reported.

Key Words : Perforating calcific elastosis, Perforating pseudoxanthoma elasticum, Periumbilical

Introduction

Pseudoxanthoma elasticum (PXE) is an inherited disorder of connective tissue in which degeneration and calcification of elastic fibers occurs in certain areas of skin, eyes and cardiovascular system. The cutaneous lesions which manifest by the 2 decade of life are characteristically confined to certain areas of stress and movement such as flexural folds of axilla, groin and popliteal fossa.¹ An acquired localised form of this disease affecting the reticular dermis of the skin in the periumbilical region of obese, middle-aged, multiparous black women has been described.² Transepidermal elimination of the altered elastic tissue through hyperkeratotic draining papules is a feature observed in both forms of this condition and was termed perforating PXE by Lund.³ The uncommon localised variant has been more descriptively designated perforating calcific elastosis.^{2,4} Trauma in some form was considered to be a predisposing factor in its causation.^{2,4} The case presented here is an unusual sequel to a scald.

Case Report

A 45-year-old multiparous woman complained of a skin lesion on the anterior abdominal wall. It had been present for 1 year and was gradually increasing in size. She had a scald at the site due to hot oil 1½ years earlier and had self medicated it with a mud pack. She had been on treatment for diabetes mellitus for 7 years. There was no history of any abdominal surgery.

On examination the patient was obese, normotensive and her fasting blood sugar was 130mg%. All other systems and laboratory investigations were normal. There was a hyperpigmented, non-pruritic, verrucous plaque in the supra umbilical region. The skin over the rest of the abdomen and body was unremarkable.

The excised specimen of skin measured 3x2 cms, had verrucous, hyperpigmented surface with multiple, discrete, keratotic, punctate papules (Fig.1). The largest papule was ulcerated. H & E stained sections showed curled, fragmented, basophilic elastic fibers in the reticular dermis. These elastic fibers were seen extruding to the surface through a tunnel in the hyperkeratotic hyperplastic epidermis. They were positive for calcium with von Kossa stain (Fig.2). They stained blue with PTAH and red with Masson's trichrome stain. Verhoeff's stain showed the elastic fibers to be short, curled

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and thick. The papillary dermis was normal. A diagnosis of perforating calcific elastosis was made.

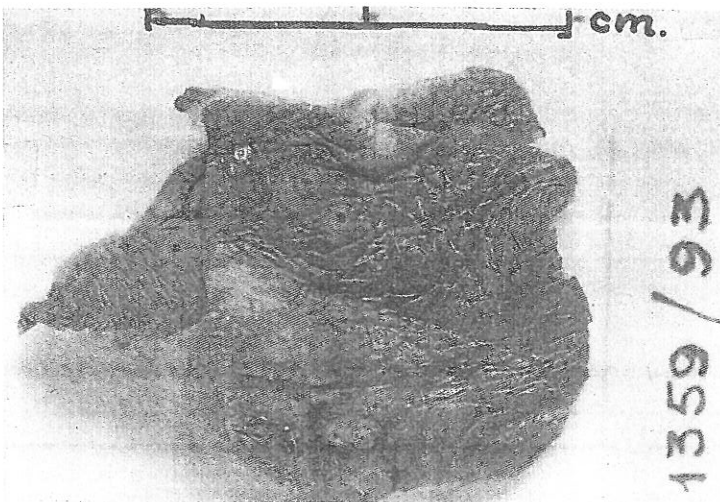


Fig. 1. Hyperpigmented skin with multiple, discrete, keratotic punctate papules

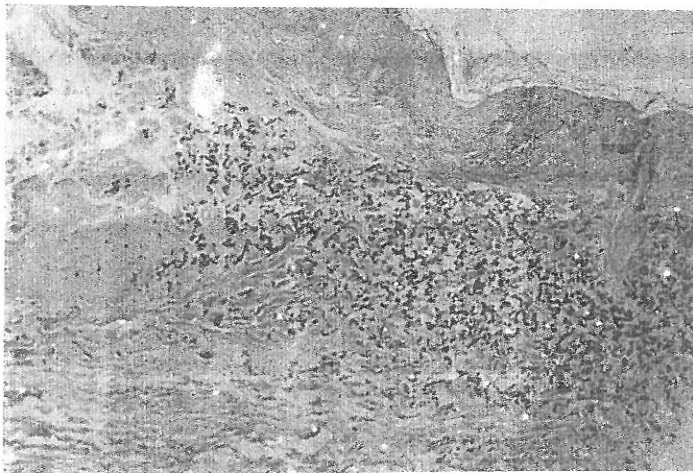


Fig. 2. Calcification of elastic fibers in mid reticular dermis with transepidermal elimination (Von Kossa stain x 160)

Discussion

A total 12 cases of periumbilical perforating pseudoxanthoma elasticum have been reported since 1979.⁵ All had a somewhat similar medical history of cardiovascular abnormalities probably related to either hypertension or diabetes mellitus.⁵ Schwartz and Richfield⁶ conclude that these patients with solitary cutaneous plaque could represent PXE affecting only one organ system and should be carefully examined for possible systemic involvement. Hicks et al² hypothesised that in genetically predisposed

individuals trauma of multiple pregnancies may cause damage to the periumbilical elastic fibers. The various forms of stresses implicated as an initiating factor include obesity, multiparity², massive ascites⁷ and surgery.^{5,8} Included in the acquired group are individuals exposed to calcium salts, including old farmers exposed to Norwegian saltpetre (calcium and ammonium nitrate).¹ One patient who was diabetic, obese and multiparous had a past history of scald in the affected area to which she had applied mud. It is surmised that trauma in the form of scald and various salts that may be present in the mud would have resulted in perforating calcific elastosis. She had no other manifestations of classic PXE. The alternative term perforating calcific elastosis has been used here to describe this condition to avoid the implications of classic PXE while retaining morphologic accuracy.

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