An unusual periumbilical plaque

A 55-year-old menopausal woman presented with an asymptomatic raised lesion around the umbilicus since the past 3 years. The lesion was gradually increasing in size. There was no history of any abdominal surgeries. The patient was hypertensive and on treatment with atenolol 50 mg OD since 4 years. She was a multiparous woman with 4 children.

On examination, the abdominal skin was loose and showed multiple striae gravidarum. The supra-umbilical area showed a well-demarcated, irregular, yellowish, crinkled, grooved plaque about 3 cm in size [Figure 1]. On palpating the lesion, there was decreased skin turgor and the skin felt inelastic.

Blood biochemistry, including lipid profile and serum calcium, was normal. Electrocardiogram, 2D-echocardiograph and ultrasonography of the abdomen and pelvis were normal. Detailed ophthalmologic examination was normal. Findings of a skin biopsy from the lesion are shown in Figures 2-3.

WHAT IS YOUR DIAGNOSIS?

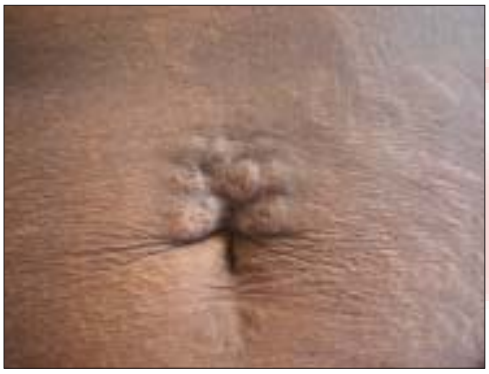

Figure 1: Close-up of well-demarcated plaque located above the umbilicus

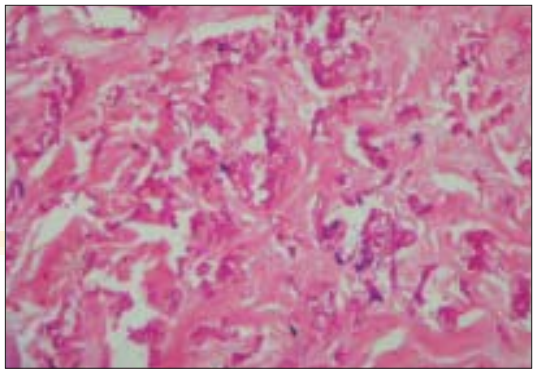

Figure 2: H and E stain (×400)

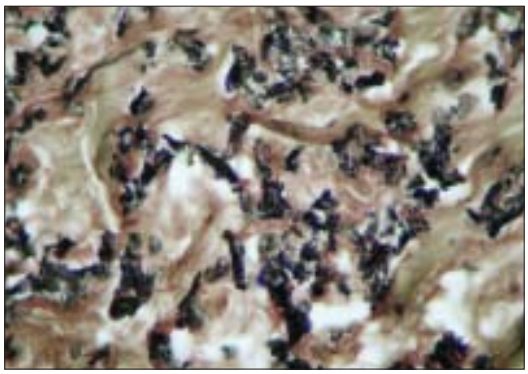

Figure 3: Verhoeff-van Gieson stain (×400)

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Diagnosis: Localized acquired periumbilical pseudoxanthoma elasticum

DISCUSSION

Hematoxylin and eosin–stained sections from the lesion showed fragmented, small, wavy, crinkled, and clumped eosinophilic elastic fibers giving a 'ravelled wool' appearance. These were confirmed to be elastic fibers as they stained black with Verhoeff-van Gieson stain [Figure 3]. Von Kossa stain was negative for calcium. The histopathology was diagnostic of pseudoxanthoma elasticum (PXE).

PXE is a rare genetic disorder characterized by progressive fragmentation of elastic fibers in the skin, the retina, and the cardiovascular system. Cutaneous lesions are usually apparent by the second decade of life and are characteristically confined to certain areas of stress and movement such as flexural folds of axilla, groin, and popliteal fossae. Small, yellow papules 1 to 5 mm in diameter are seen in a linear or reticular pattern and may coalesce to form plaques. The skin takes on a 'plucked chicken,' 'Moroccan leather,' or cobblestone appearance.[1] Skin biopsy shows fragmented, swollen, degenerate, and clumped elastic fibers in the middle and deep reticular dermis with characteristic 'ravelled wool' appearance. Collagen fibers are also abnormally split into small fibers. Special stains for elastic fibers (e.g., Verhoeff van Gieson, Orcein) and calcium deposits (e.g., von Kossa) can confirm the diagnosis.[2]

The acquired localized form of PXE is a disease affecting the reticular dermis of the skin in the periumbilical region of obese, middle-aged, multiparous women.[3] It was first differentiated from elastosis perforans serpiginosa on histological grounds by Lund and Gilbert in 1976.[4] The cause is as yet unknown; however, various forms of stresses on the abdominal wall connective tissue have been implicated as an initiating factor. These include obesity, multiparity, massive ascites, and abdominal surgery.[3] Association with chronic renal failure has been noted with apparent regression of skin lesions after hemodialysis.[4]

Localized acquired cutaneous pseudoxanthoma elasticum is a new designation proposed for this nonheritable form of pseudoxanthoma elasticum (PXE). The skin lesions are clinically, histologically, and ultrastructurally similar to those seen in the inherited type. It is differentiated from the classical syndrome by late onset, absence of flexural clinical lesions, negative family history, and no systemic manifestations. Ophthalmologic examination has revealed angioid streaks in some cases; however the other classical systemic manifestations of PXE are absent.[4]

The onset of the disease is in the fifth to sixth decade of life in perimenopausal women. The cutaneous lesions are well-demarcated asymptomatic periumbilical plaques with keratotic papules at their periphery. Most reported cases have been hypertensive[4] and hypercalcemic. The histology reveals morphologically altered elastic fibers throughout the dermis. These may be calcified and also may extrude through the skin surface by transepidermal elimination, simulating elastosis perforans serpiginosa. However, the elastic fibers are situated only in the upper dermis, and are coarser, straighter, and more eosinophilic in the latter as compared to fragmented, clumped, and calcified elastic fibers throughout the mid and deep dermis in PXE.

Localized periumbilical PXE is rare, and approximately 20 cases have been reported in literature till date.[4] There is currently no effective treatment for the skin lesions, but the appearance may be improved by plastic surgery. In our case, the patient was explained about the nature of the condition, and she chose to leave it untreated.

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