Case Reports

PERIUMBILICAL PSEUDOXANTHOMA ELASTICUM

V Vishalakshi, RG Torsekar

Pseudoxanthoma elasticum is an inherited connective tissue disorder affecting skin, ocular and vascular systems. We report an unusual case with cutaneous lesions simulating PXE localized to the periumbilical region with no systemic involvement. Two entities having localized PXE- like skin changes namely perforating calcific elastosis and papillary dermal elastolysis are discussed.

Key Words : Pseudoxanthoma elasticum (PXE), PXE- like papillary dermal elastolysis (PDE)

Introduction

Pseudoxanthoma elasticum (PXE) is an inherited connective tissue disorder affecting skin, ocular and cardiovascular systems.[1] Cutaneous lesions are characteristic and have been described as cobble stone , Morrocan leather or chicken appearance. Angioid streak is the hallmark of ocular involvement and occlusive vascular disease is responsible for systemic complications. Acquired and localized forms of PXE are uncommonly encountered.

Case Report

A 50- year- old woman presented with a minimally pruritic hyperpigmented lesion on the abdomen since 2 years. No relief was obtained with multiple topical and systemic antifungal/ steroid therapy. She had no lesions elsewhere and no systemic complaints. An enquiry into the obstetric history revealed that she had 3 full - term normal deliveries, no abortions and was menopausal since the past 3 years.

Cutaneous examination revealed a well-defined, infiltrated hyperpigmented plaque in the periumbilical region with cobblestoning appearance

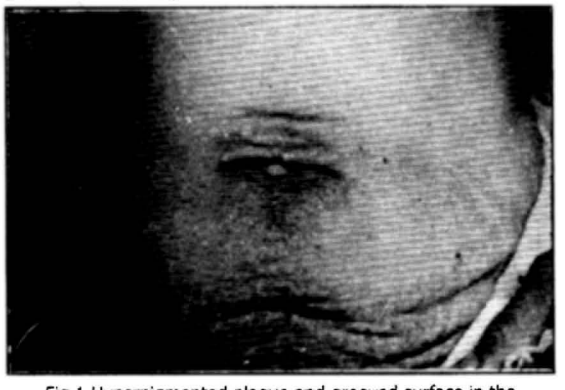

Fig.1 Hyperpigmented plaque and grooved surface in the periumbilical region and striae distensae.

at places (Fig. 1). Striae distensae was present. Mucosae were normal and no cutaneous abnormality was seen elsewhere. Systemic examination was normal.

A differential diagnosis of a localized infiltrative disorder like mucinosis or PXE was considered.

From the Department of Dermatology, CSM Hospital & Rajiv Gandhi Medical College, Kalwa, Thane, Maharashtra.
Address correspondence to:
Dr. Vishalakshi V
18, Sri Valli, Rifle Range, Ghatkoper (W) Bombay- 400 086.

Biopsy of the skin lesion revealed a hyperpigmented epidermis with clumped eosinophilic material in the mid dermis giving the characteristic revelled wool appearance suggestive of PXE (Fig 2). Von Kossa stain was positive.

A detailed systemic work up did not show any evidence of pseudoxanthomatous changes elsewhere. Routine biochemical investigations including serum levels of calcium were within normal limits.

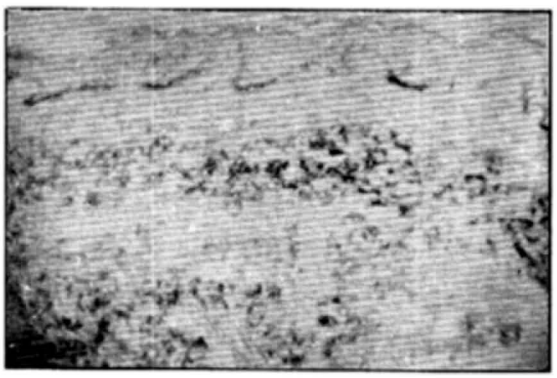

Fig.2. Histopathology: Positive Von Kossa stain showing classical revelled wool appearance.

X- ray of the chest, USG abdomen, ECG and a detailed ophthalmic evaluation did not reveal any abnormality.

Discussion

Pseudoxanthoma elasticum is an inherited disorder of connective tissue that is characterised by calcification of elastic fibres with associated anomalies of skin, ocular and cardiovascular system. Drug induced PXE- like skin changes have been described with d- penicillamine, but no systemic involvement is seen.[2]

Localized cutaneous features of PXE with no systemic involvement or prior drug intake is seen in two entities namely a) PXE - like papillary dermal elastolysis (PDE), b) Perforating calcific elastosis. Both these acquired conditions closely resemble PXE with certain variations in clinical and histological picture. PDE, a recently described entity, is a rare acquired idiopathic non- inflammatory elastolytic disorder described in aged females, clinically characterised by multiple asymptomatic slowly progressing coalescing skin-coloured papules on the neck, supraclavicular region and abdomen.[3] Histologically total loss of elastic fibres in the papillary dermis is seen. PDE probably represents a clinico-pathological pattern of intrinsic ageing.

Perforating PXE, or perforating calcific elastosis is a localized acquired defect of elastic tissue seen in obese, multiparous black women.[4] Clinically, the lesions are characterised by a well- demarcated slowly prgressive hyperpigmented plaque and the surface may be atrophic, grooved, fissured or verrucous with discharge from the edge of the lesion. Histologically PXE changes are seen associated with transepithelial elimination of calcific material, hence the name perforating calcific elastosis. The localized defect in elastic tissue occurring in the periumbilical region in multiparous women may be a response to repeated cutaneous stretching in successive pregnancies. Case reports of perforating PXE have been described either as a localized pathology or in association with hereditary PXE.[5] Our case corroborates with perforating PXE, however transepithelial elimination was not observed, which may occur with due course of time. This case has been reported for its rarity.

References

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