Dermoscopic Intrusion into a Pathological Extrusion

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Clinical Presentation

A 59-year-old multiparous obese woman presented with asymptomatic dark lesions in the periumbilical region for the past 2 years. There was no history of chest pain, abdominal pain, intermittent claudication, and visual disturbances. No history of similar complaints among the family members. On local examination, the patient had a few hyperpigmented keratotic papules with surrounding atrophic depressed plaques in a background of brownish-yellow pigmentation in the periumbilical region as shown in Figure 1. Findings observed in the dermoscopic examination (polarized mode, Dermlite DL 4, 10x magnification) are shown in Figure 2a and b. An ophthalmological examination was done for the patient, which was found to be normal. Complete hemogram, blood sugar, urine routine, serum calcium and phosphorous levels, renal and liver function tests, and electrocardiogram were normal. Histopathological examination of elliptical incisional skin biopsy was done from one of the hyperkeratotic papules [Figures 3, 4a and b]. The patient was treated with 0.05% tretinoin cream and topical 0.1% mometasone furoate cream and was advised to follow-up.

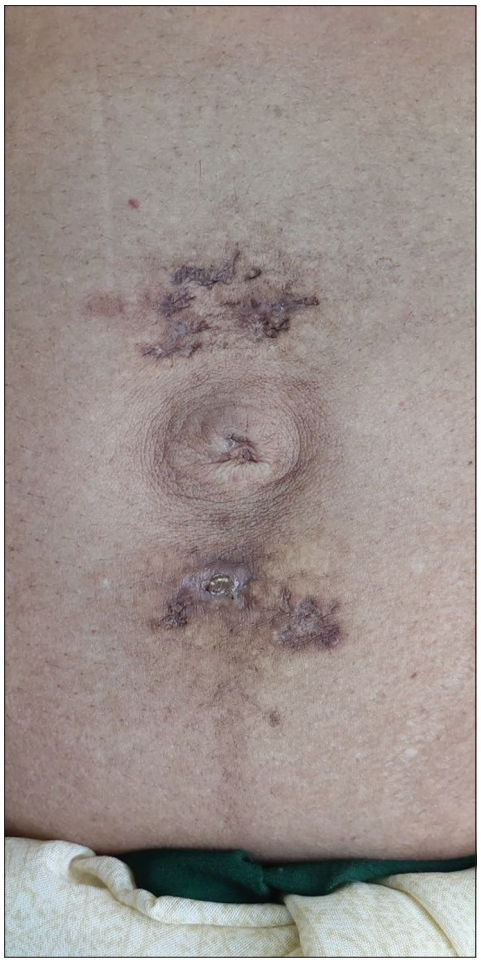

Figure 1: Few hyperpigmented keratotic papules with surrounding atrophic depressed plaques in a background of brownish-yellow pigmentation in the periumbilical region

What is your diagnosis?

Diagnosis

Periumbilical perforating pseudoxanthoma elasticum (Perforating calcific elastosis).

Discussion

Pseudoxanthoma elasticum (PXE) is an inherited elastic tissue disorder characterized by elastic fiber degeneration and calcification in the cutaneous, ocular, and cardiovascular systems. Whereas periumbilical perforating pseudoxanthoma elasticum (PPPXE) is considered a rare acquired atypical perforating disease characterized by periumbilical plaques with reticulated, atrophic, ridged, fissured, or verrucous surfaces.[1] It is also characterized by degeneration, calcification, and transepidermal elimination of elastic fibers.[2] Hence, the term perforating calcific elastosis was coined by Lever and Schaumberg.[3] Perforating PXE has also been reported in the periareolar region.

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Risk factors of PPPXE include middle age, obesity, multiparity, and the black population.[1,3] Excessive abdominal wall stretching in the form of multiple pregnancies, massive ascites, and multiple abdominal surgeries are also frequent associations.[3] Abdominal trauma due to excessive stretching leads to degeneration and fracture of elastic fibers which subsequently get mineralized with calcium during the reparative process.[4]

Until 1976, it was thought to be a disease coexisting with elastosis perforans serpiginosa, when Lund and Gilbert proved it to be a separate entity and termed it as perforating pseudoxanthoma elasticum.[4] Still, there is a high degree of dilemma about whether PPPXE is a distinct acquired entity or a localized cutaneous form of hereditary PXE, owing to the occurrence of angioid streaks in a few patients of PPPXE. Although there are reports of associations of angioid streaks in PPPXE, neither of them was diagnostic of PXE retinopathy.[1,4]

The common dermoscopic findings observed in available reports are yellow-brown areas, arcuate/curvilinear hyperpigmented lines, telangiectasia, hairpin and dotted vessels, and keratotic plug.[2] The arcuate hyperpigmented lines corresponded to altered elastic fibers or calcific deposits, white halo with pigment loss, and erythema with inflammatory infiltrates. The keratotic plug in a lamellated pattern with surrounding collarette scales over dam-shaped uplift was congruous with the transepidermal elimination of elastic fibers with the resultant cup-shaped depression of the epidermis.[2]

The differential diagnoses for PPPXE include reactive perforating collagenosis, Kyrle's disease, elastosis perforans serpiginosa, and perforating granuloma annulare. The comparison of dermoscopic features of various perforating dermatoses is tabulated in Table 1, which can help to differentiate PPPXE from other perforating disorders.[5-10]

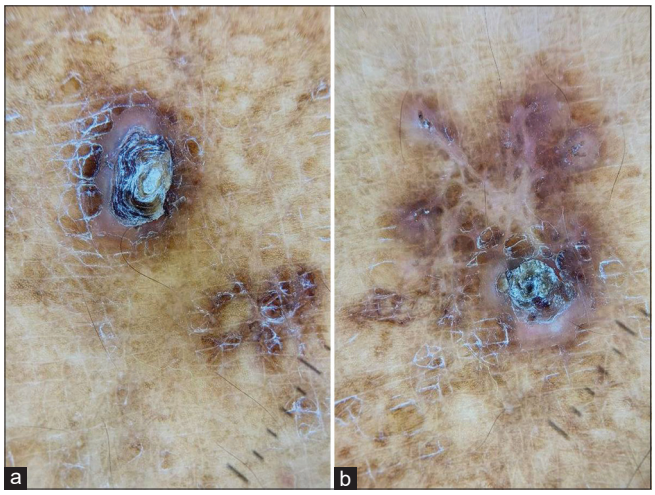

Figure 2: (a and b) Dermoscopic examination (polarised mode, Dermlite DL4, 10x) showing yellowish-brown structureless areas with arcuate hyperpigmented lines, white halo, surrounding erythema and atrophy, keratotic plug with collarette, and lamellated pattern of scales over a dam-shaped uplift

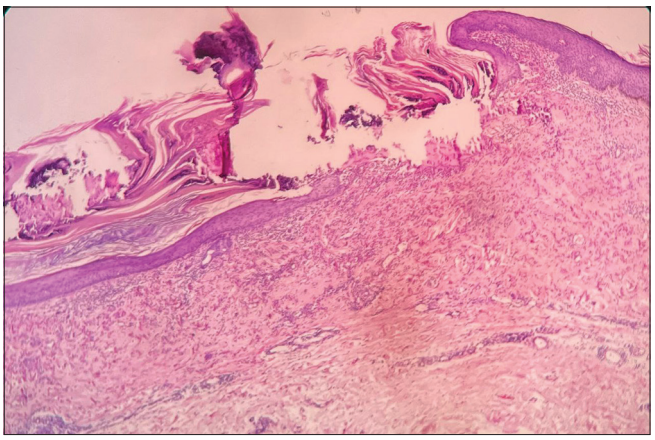

Figure 3: Orthokeratosis, mild acanthosis, and extrusion of fragmented elastic fibers through the tunnel in the epidermis ending in a keratin-filled crater (H&E, 10x)

Table 1: Comparison of dermoscopic features of various perforating dermatoses

Reactive perforating collagenosis[5]	Elastosis perforans serpiginosa[6,7]	Kyrle's disease[8]	Acquired perforating dermatosis[9]	Periumbilical perforating pseudoxanthoma elasticum[10]
• Central yellow-brown structureless area • Whitish ring • Pink white halo	• Central white or yellow structureless area • Arborizing linear vessels • Brownish-crusted papules in the periphery • Surrounding white halo (archipelago pattern)	• Central crust surrounded by keratotic scales • Structureless white-grey areas • Structureless pink area • Structureless brown area with peripheral scale	• Yellow-brown homogenous structureless area in the center • White irregular ring • Dotted and linear vessels distributed radially • Uplift at the periphery of the lesion	• Yellow-brown structureless area with arcuate hyperpigmented lines • White halo • Keratotic plug with surrounding collarette of scaling • Lamellated pattern of scales over a dam-shaped uplift • Erythema with areas of atrophy • Telangiectasia • Hairpin and dotted vessels

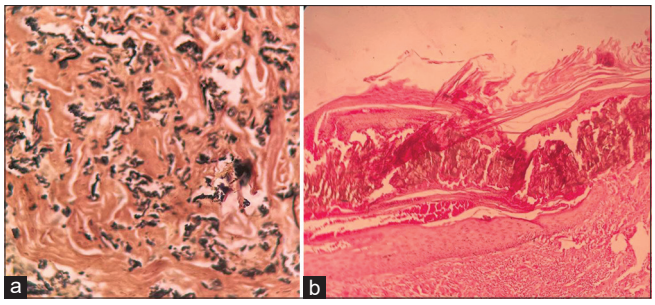

Figure 4: (a) Verhoeff Van Gieson stain (40×) showing irregular, clumped fragmented elastic fibers (b) Von Kossa stain (10×) showing areas of calcification

There are not many reports available describing the treatment options for PPPXE. Some have tried dietary calcium restriction to <800 mg/day and phosphate binders which were effective in some cases.[11] Other treatments like topical steroids have also been found to be ineffective. Reconstructive surgeries have also been suggested as a temporary relief for prominent skin redundancy.[12] It is necessary for clinicians to appreciate that although PPPXE shares some features with PXE, the former is acquired and localized only to the abdominal skin.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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