A Puzzling Periumbilical Pigmented Plaque

Question
A 76-year-old female came to us for evaluation of gradually progressing hyperpigmented papules over her abdomen of 10 years duration. There was occasional discharge and few lesions healed spontaneously with scarring and hyperpigmentation. She reported minimal improvement with topical tretinoin 0.05% used previously. Her medical history included tuberculous ascites treated 11 years back, diabetes with uncontrolled blood sugars, chronic liver disease secondary to non-alcoholic fatty liver disease, complete heart block with a permanent pacemaker implanted two years back. She has nine children, the last childbirth being 35 years back, and had not undergone any surgeries. Her family history was unremarkable. Her body mass index was 21.8 and systemic examination was normal. There were hyperpigmented atrophic plaques studded with multiple hyperpigmented keratotic papules with central crusting and few atrophic scars in the periumbilical region [Figure 1].

Histopathological examination of the hyperkeratotic papule showed intracorneal necrotic cellular and basophilic debris with mild acanthosis [Figure 2c]. Short, frayed, basophilic fibers were seen in the mid and deep-dermis [Figure 2a and b]. These abnormal fibers were demonstrated by orcein stain [Figure 3a] and negative for Masson trichrome stain. Orcein also highlighted transepidermally eliminated abnormal fibers within the stratum corneum. Von Kossa staining showed scattered coarse black stippling in the dermis [Figure 3b].

What is your diagnosis?

Diagnosis: Periumbilical perforating pseudoxanthoma elasticum.

Discussion
Periumbilical perforating pseudoxanthoma elasticum (PPPXE) is an acquired perforating dermatosis (APD) classically presenting as asymptomatic or mildly pruritic hyperpigmented atrophic plaques with scaly raised serpiginous margins, studded with discrete and coalescent keratotic papules over the abdomen. Although it is usually seen in middle-aged, obese, multiparous women of African-American origin, cases have been reported in other races and a single case reported in a nulliparous woman.[1,4] The etiopathogenesis of PPPXE is unclear. While some believe it to be a localized cutaneous feature of hereditary pseudoxanthoma elasticum (PXE), some consider it an acquired distinct clinicopathological entity.[2] It was first described by Hicks et al.[1] in six African-American women and he proposed that this condition is seen in women genetically predisposed to PXE, with the delayed onset explained by the greater resistance of heavily pigmented skin to dystrophic changes of PXE. Lending credence to this theory is the co-existence of hypertension, angioid streaks, and other flexural involvement in some cases.[2,4] Nevertheless, the onset of lesions in the 5th-6th decade, lack of other flexural involvement, systemic symptoms, and family history were consistent with its acquired nature in our patient. It is hypothesized that the mechanical stress to the abdominal skin secondary to multiparity, ascites, surgery, trauma, obesity causes degeneration and calcification of elastin in the dermis, which undergoes transepidermal elimination.[2,4] Deranged calcium-phosphate metabolism in chronic renal failure (CRF) can also lead to similar elastin bioneralization.[2,4] Although our patient was not obese, the multiple

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pregnancies and ascites in the past could have triggered the condition.

On histopathology, short, thickened, frayed, and fragmented elastin fibers are seen predominantly in the mid-dermis. The papillary dermis is usually unaffected, except at the sites of transepidermal elimination. Calcification of the abnormal elastin fibers imparts a basophilic appearance and accounts for the black staining with von Kossa stain.\(^{[2,4,5]}\) Elastosis perforans serpiginosa (EPS) is a close clinical and histological differential; however, the thickened elastin fibers are non-calcified, more eosinophilic, and predominantly located in the papillary dermis in EPS.\(^{[2,4,5]}\)

Though topical retinoids or steroids have been tried, no significant benefit has been reported.\(^{[4]}\) In CRF, the skin lesions may regress with hemodialysis.\(^{[5]}\)

Perforating calcific elastosis would be a more appropriate term to describe this rare APD seen in obese, multiparous females, which is histopathologically similar to PXE, but without its extracutaneous features.\(^{[2,5]}\)

**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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