Dermoscopy of Pseudoxanthoma Elasticum

Sir,

Pseudoxanthoma elasticum (PXE), also known as Gronblad-Strandberg syndrome, is an inherited rare disorder characterized by the aberrant mineralization of soft tissue and the fragmentation of elastic fibers, which results in multiorgan disorder primarily involving the skin, eyes, and cardiovascular system.[1] The basic defect in PXE lies on the short arm of chromosome 16p13, and it is caused by mutation in ABCC6 (ATP-binding cassette) gene.[2]

A 21-year-old woman presented with complaints of asymptomatic yellowish raised lesions over sides of neck and axilla, gradually progressing since 7 years. No similar complaints were present in family members. General and systemic examination was normal. Cutaneous examination revealed multiple symmetrically distributed, confluent yellowish papules arranged in a linear and reticulate pattern coalescing to form plaques, giving it a cobblestone appearance, present over lateral and anterior aspects of neck and axilla [Figure 1]. The rest cutaneous examination was normal.

The dermoscopy of neck lesions was performed using 3Gen DermLite DL4 (CA, USA) in a 10× polarized mode, which revealed multiple irregular yellowish-white areas (clods) arranged in a linear and reticulate manner alternating with linear vessels on a light red background [Figure 2a and b]. On fundoscopy, *peau d'orange* appearance was seen. The rest of investigations were normal. Histopathology revealed faintly basophilic distorted (curled and frayed) elastic fibers in mid-reticular dermis. Orcein staining showed deep black colored elastic fibers with characteristic elastorrhexis. von Kossa stain confirmed the deposition of calcium in dermis [Figure 3a–f]. On the basis of aforementioned findings, a final diagnosis of PXE was reached.

Figure 1: Multiple symmetrically distributed, confluent yellowish papules arranged in a linear and reticulate pattern coalescing to form plaques, with cobblestone appearance over lateral and anterior aspects of neck and axilla

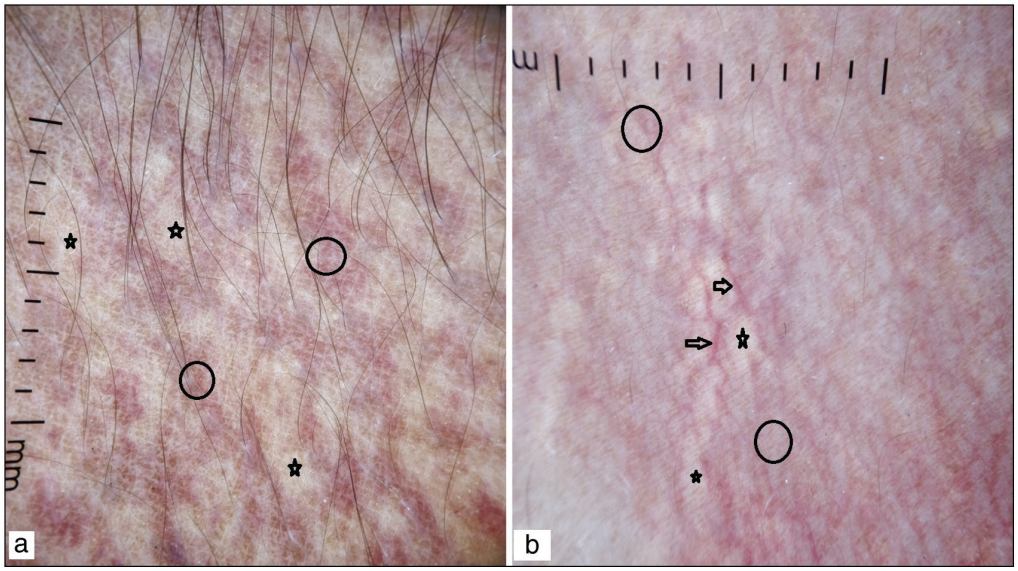

Figure 2: (a) and (b) Dermoscopy of neck lesions revealed multiple irregular, yellowish-white areas/clods (black star) arranged in a linear and reticulate manner alternating with linear vessels (black arrow) on a light red background (black circle) ([a and b] dermoscopy, 10× polarized mode)

Table 1: Review of literature of dermoscopy of PXE

S. no.	Authors	Year	Age/sex	Dermoscopic description
1	Lacarrubba *et al.*[3]	2017	9/F 38/F	Multiple irregular, yellowish areas coalescing to form linear, parallel strands in one case. Multiple blotchy, yellowish areas alternating with prominent superficial linear vessels in the second case.
2	Singh *et al.*[5]	2017	22/F	Multiple irregular, yellowish areas arranged as linear strands alternating with multiple linear vessels.
3	Kawashima *et al.*[4]	2018	74/M 72/F	Coalescing and reticulated yellowish-white clods on a light purplish-red background and subtle reticulated vessels in one case. Yellowish-white clods on a light purplish-red background with a lack of vessel findings in the second case.
4	Salas-Alanis *et al.*[6]	2019	56/F	Multiple irregular, yellowish-white areas alternating with prominent, superficial linear vessels, on a light purplish-red background.
5	Chauhan *et al.*[7]	2021	20/F	Multiple yellowish-white structures (black arrow) coalescing irregularly to form linear streaks, interspersed with areas of ill-defined faint erythema.
6	Our case	2022	21/F	Multiple irregular, yellowish-white areas (clods) arranged in a linear and reticulate manner alternating with linear vessels on a light red background.

The first dermoscopic description of PXE was given by Lacarrubba *et al.* in 2017, in which multiple irregular, yellowish areas with prominent linear vessels are depicted.[3] The yellowish white clods in PXE correspond to elastolysis or calcium deposition in mid-dermis. Purplish-red background in PXE is due to mid-dermal vasodilation. The prominent superficial linear vessels may be caused by vascular rearrangement with underlying dermal elastolysis.[4] Dermoscopy may help differentiate PXE and PXE-like papillary dermal elastolysis on the basis of color of clods and background. In the former, yellowish-white clods with light purplish red background is seen, and light-yellow clods with flesh colored background is seen in the latter.[4] Thus, dermoscopy could be useful for non-invasive early diagnosis of skin lesions of PXE. There are very few reports describing the dermoscopy of PXE in the literature [Table 1]. We report this case because of its rarity in the Indian literature.

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.

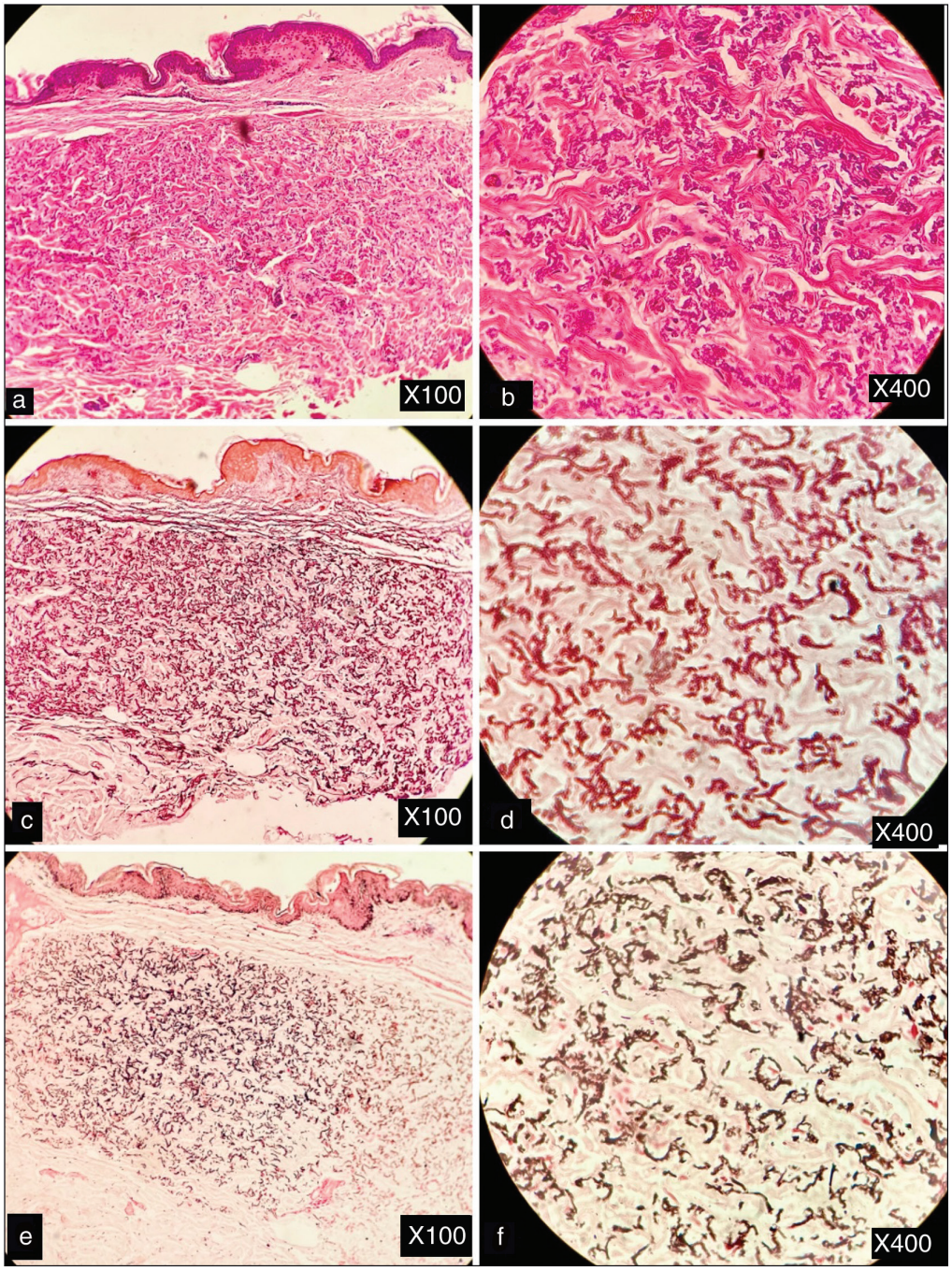

Figure 3: (a–f) Histopathology revealed faintly basophilic distorted (curled and frayed) elastic fibers in mid-reticular dermis (Hematoxylin-eosin stain; original magnifications: a, X100; b, X400;); Orcein staining showed deep black colored elastic fibers with characteristic elastorrhexis (Orcein stain; original magnifications: c, x100; d, X400); von Kossa stain confirmed the deposition of calcium in dermis (Von Kossa stain; original magnification: e, X100; f, X400).

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