

A rare case of *Angiokeratoma circumscriptum naeviforme* on trunk: through the lens

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Introduction

Angiokeratomas are rare vascular lesions characterised by papillary dermal blood vessels with secondary epidermal changes.

Angiokeratoma circumscriptum naeviforme (ACN) is the rarest clinical variant of angiokeratoma which often presents at birth or in infancy in a segmental and/or zosteriform pattern as a result of a nevoid vascular malformation.¹

Case report

History

- A 14-year-old boy with gradually progressive blue to black linear lesions over his left flank, since 2 years age.
- The lesions were asymptomatic but with episodes of bleeding post trauma.

Examination

- Multiple dark purple to blue well-circumscribed discrete to confluent verrucous papules and plaque in a blaschkoid distribution over left flank (figure1).
- These were firm to hard in consistency, indurated, non-tender and non-pulsatile to touch.
- Regional lymph nodes were non-palpable.

Investigations

Dermoscopy

- well demarcated areas of deep red to blue and violaceous round to oval lacunae and whitish veil over few areas (figure 2).

fig 1



Fig 2

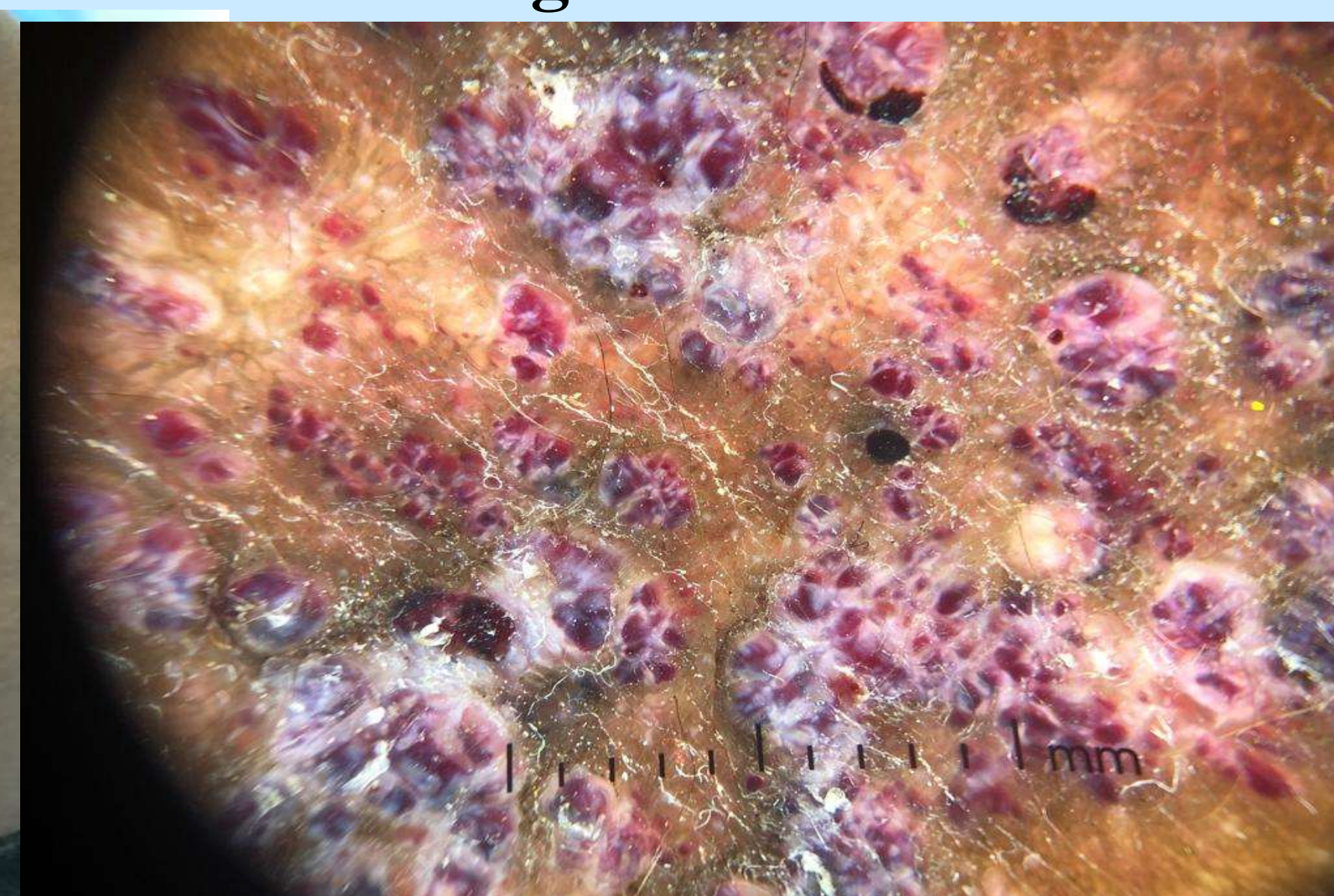


figure 3a

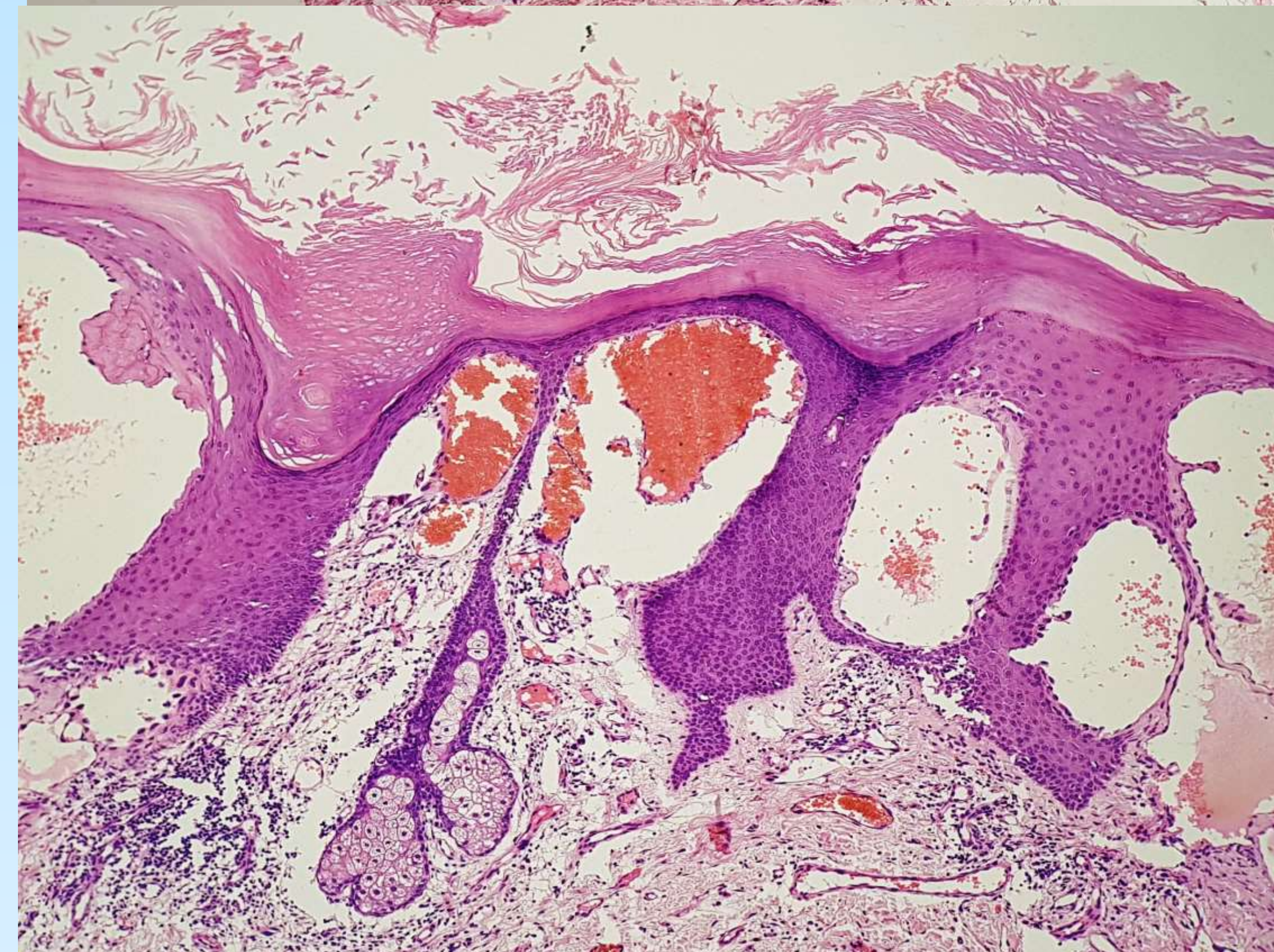
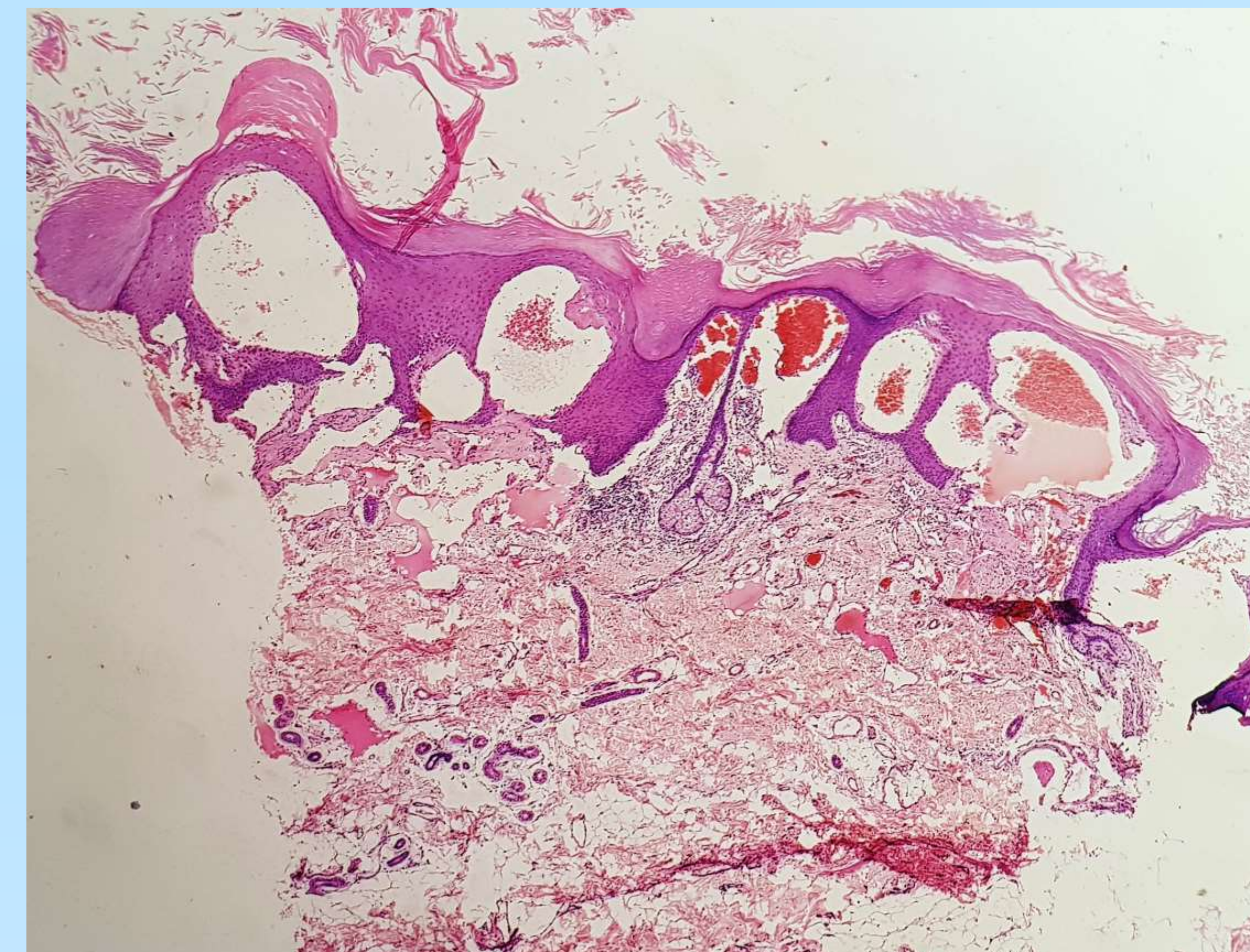


Figure 3b

Histopathology

Hyperkeratosis, mild papillomatosis and irregular acanthosis of the epidermis with elongation of the rete pegs encircling the dilated vascular spaces in the papillary dermis.

Vascular ectasia appears to extend into the epidermis. Moderate perivascular inflammation was seen in upper dermis. (H&E, 100X) (figure 3a). No vascular ectasia was found in the reticular dermis or in the subcutis.

High power view revealed a vascular lesion in the superficial dermis extending into the epidermis. The vascular spaces were engorged with red blood cells. (H&E, 200X) (figure3b).

Discussion

- There are only a few reports of ACN in the world literature.
- ACN has so far not been reported to afflict trunk.
- Considering clinical similarity of ACN and verrucous hemangioma, histopathological examination is imperative for diagnostic confirmation.
- dermatoscopy being non-invasive and novel tool can aid in establishing the diagnosis and exclusion of other pigmented lesions like malignant skin tumors, melanocytic nevi and vascular lesions such as verrucous hemangioma, angiomas, and pyogenic granulomas.
- Verrucous hemangioma though showing similar pattern as ACN on dermatoscopy lack well circumscribed lacunae.⁵

References

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- Ghosh SK, Bandyopadhyay D, Ghoshal L, Haldar S. Angiokeratoma circumscriptum naeviforme: a case report of a rare disease. *Dermatol Online J.* 2011; 11:15-7.