Unilateral blaschkoid pseudokaposi's sarcoma: through the lens

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Introduction

- Acroangiodermatitis/pseudokaposi's sarcoma is a reactive cutaneous angiodysplasia ascribed to increased venous pressure and stasis caused by venous insufficiency, arteriovenous malformations, arteriovenous shunts and lower limb paralysis or amputation stumps.
- Lesions along Blaschko's lines have not been described so far.

Case report

- A 17-year-old girl with asymptomatic dark brown to purple papules and plaques over the left leg of 6 years duration.
- Swelling of the left foot and ankle.
- Examination: brown to violaceous ill to well defined papules and plaques arranged in a blaschkoid distribution over left leg extending from medial malleolus to medial aspect of thigh above the knee (fig 1).

Investigations

- HIV testing by ELISA was negative.
- Color doppler imaging and CT angiogram of left lower limb revealed multiple dilated tortuous superficial veins below knee up to ankle along with incompetence of sapheno-popliteal junction (fig 2)

Histopathology

- hyperkeratosis and irregular acanthosis of the epidermis. Extravasated erythrocytes and a sparse perivascular mononuclear cell infiltrate admixed with occasional eosinophils (fig 3a H&E, 100X)
- Papillary and reticular dermis showed lobular proliferation of small capillaries in a loose stroma. Blood vessels in the dermis were lined by plump endothelial cells without any atypia. (fig 3b) (H&E, 400X)
- Hemosiderin deposits. (fig 3c Perl's stain 100 x)
- CD 34 positivity was observed in endothelial cells of hyperplastic vessels. (fig 3d)

fig 1

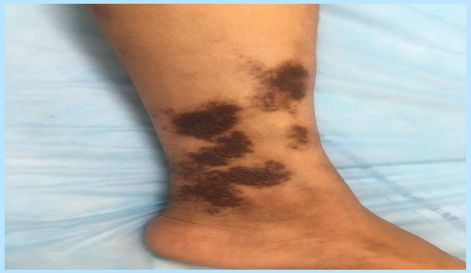


fig 3a

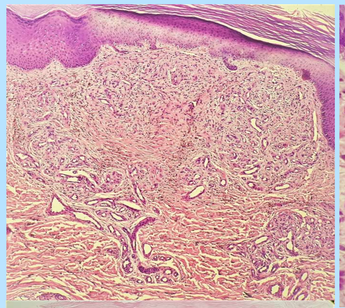


fig 3b

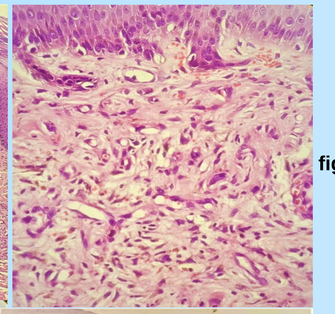


fig 3c

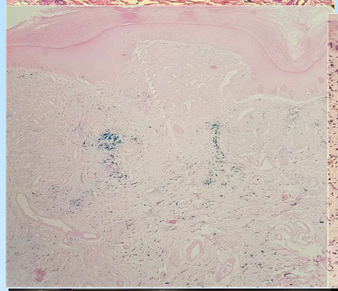


fig 3d

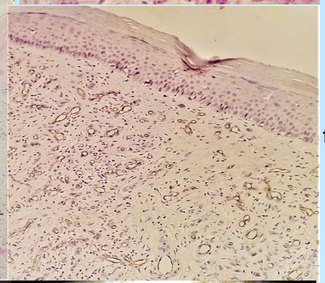


fig2

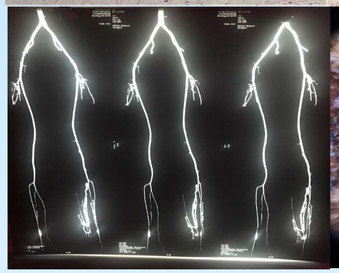
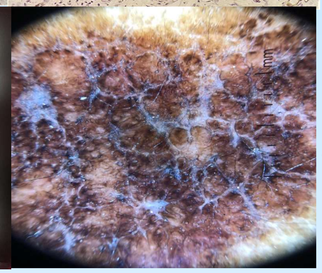


fig 4



Dermatoscopy

Blue lacunae, whitish veil, white rail lines, prominent vascularisation and brownish globules. (fig 4)

Discussion

- Somatic mutation might have resulted in abnormal differentiation, migration and placement of vascular endothelial cells in skin.
- Diagnosis of acroangiodermatitis is confirmed by its characteristic histopthological feature.
- Further, absence of atypia, promontory sign, low density of inflammatory infiltrate.
- CD 34 positivity localised to plump endothelial cells while sparing the surrounding stroma differentiate acroangiodermatitis from Kaposi's sarcoma.

Reference

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2. Kanitakis J, Narvaez D, Claudy A. Expression of the CD34 antigen distinguishes Kaposi's sarcoma from pseudo-Kaposi's sarcoma (acroangiodermatitis). Br J Dermatol. 1996;134:44-6.