

## CASE REPORTS

### SPONTANEOUS BLEEDING, CORNU CUTANEUM, NEVUS FLAMMEUS IN ANGIOKERATOMA CIRCUMSCRIPTUM

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A case of angiokeratoma circumscriptum of the upper limb with extensive lesions, spontaneous bleeding during episodes of fever, cornu cutaneum and nevus flammeus is reported.

**Key words:** Angiokeratoma circumscriptum, Nevus flammeus, Cornu cutaneum, Association.

Angiokeratoma circumscriptum (AC) is a superficial haemangioma or telangiectasia associated with reactive hyperkeratosis of the epidermis, occasionally involving the arms.<sup>1</sup> Association with nevus flammeus is reported.<sup>2</sup> Cornu cutaneum known to occur in cavernous haemangioma<sup>3</sup> has not been reported in AC. Our case with extensive AC, grossly deforming the right forearm and hand, was accompanied by nevus flammeus, cornu cutaneum and inexplicable spontaneous bleeding from lesions during episodes of fever.

#### Case Report

An 8-year-old boy presented with multiple asymptomatic swellings over right upper limb present since birth. The lesions were gradually increasing in size. Episodes of fever associated with spontaneous bleeding occurred at weekly intervals. There was no history of any bleeding diathesis or chills and rigor accompanying the episodes of fever.

Examination revealed blue-black

irregularly distributed swellings over the right forearm, hand and palm (Fig. 1). Lesions, 2 to 4 cm in size, were soft to firm in consistency. Marked hyperkeratosis (the mass height exceeding the diameter) was seen over two swellings. A port-wine stain was seen over the middle third of right forearm. Length of the limb was unaltered. Neurological examination revealed grade IV motor power of right forearm muscles. Systemic examination did not reveal any abnormality.

Haemoglobin, TLC, DLC, platelet count, BT, CT, prothrombin time and ESR were nor-

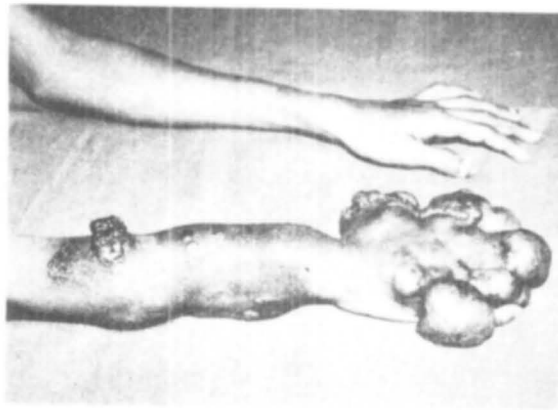


Fig. 1. Angiokeratoma circumscriptum involving the right forearm and hand.

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mal. Occult blood was not detected in stools. X-rays of chest and upper limb were normal. Excision biopsy of a lesion revealed hyperkeratosis, papillomatosis and greatly dilated capillary spaces enclosed in the epidermis suggestive of AC. Most of the lesions were excised under general anaesthesia followed by skin grafting. Recovery was uneventful, however the child was lost for follow-up.

### Comments

AC is a malformation of the vessels of papillary dermis,<sup>1</sup> usually present at birth.<sup>4</sup> The majority occur on lower leg and foot, they are less common on the thigh and buttock and can occasionally involve the arms, trunk and penis.<sup>1</sup> Association of AC with nevus flammeus,<sup>2</sup> cavernous haemangioma,<sup>3</sup> Klippel-Trenaunay syndrome<sup>5</sup> is reported. Cutaneous horn not yet reported in angiokeratomas,

extensive involvement of the upper limb (an uncommon site), inexplicable spontaneous bleeding during episodes of fever and nevus flammeus were the unusual features in our patient.

### References

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