A kaposiform haemangioendothelioma successfully treated with topical tacrolimus and compression bandaging

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Case History

- A male term infant presented at birth with a large tumour on his right forearm measuring 5 x 4 x 3cm (Figure 1).
- The lesion was firm non-pulsatile and purpuric, with coarse surface telangiectasia.
- The baby was systemically well and clinical examination was otherwise normal.
- Ultrasound and MR imaging showed a high-flow vascular lesion confined to the subcutis with no muscular invasion.
- Histologically this was a GLUT1-negative vascular tumour composed of spindloid and glomeruloid nodules consistent with a Kaposiform haemangioendothelioma (KHE).
- D-Dimers were elevated (6.22ng/ml).
- Fibrinogen and platelet count were normal.

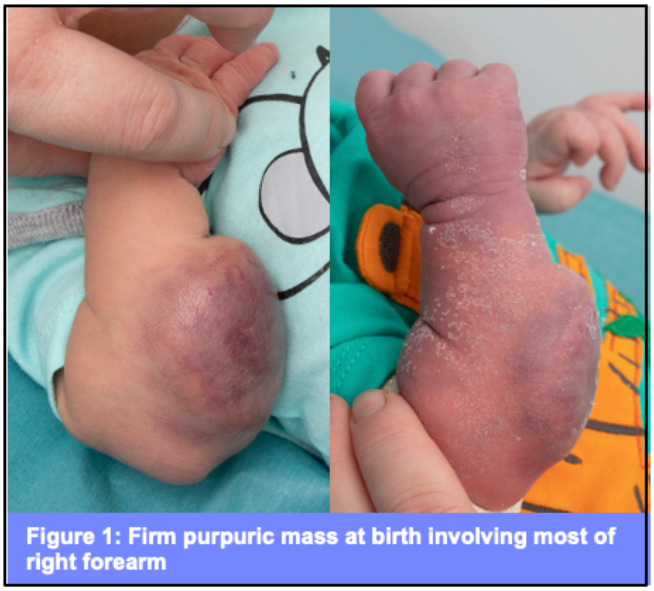

Figure 1: Firm purpuric mass at birth involving most of right forearm

Clinical Course

- In the absence of coagulopathy, a conservative approach was taken.
- Twice daily topical tacrolimus 0.1% was introduced at week 2 of life with light compression bandaging (double layer of Tubifast bandage).
- Noticeable improvement was present after just one week of treatment.
- Over the following 6 months, the lesion dramatically lightened and completely flattened. (Figure 2)
- D-dimers gradually reduced and normalised by 3 months of age.
- Compression bandaging was stopped after 6 months, with no rebound growth of the KHE.

Discussion

- KHE is a rare potentially life-threatening vascular tumour.
- Typically it presents as a solitary indurated soft tissue mass on the head, neck or extremities, but lesions range widely from small, superficial nodules or plaques to large, deep infiltrative tumours with associated coagulopathy.
- The histological hallmark of KHE are infiltrating, defined, rounded, and confluent nodules composed of spindle and endothelial cells.[1]
- Significant morbidity and mortality can occur due to local invasion/compressive effects, high-output heart failure or development of Kasabach-Merritt Phenomenon.[1]
- Therapeutic options include high dose corticosteroids, antiplatelet agents, sirolimus or vincristine, often used in combination.

Figure 2: Complete flattening and resolution of the KHE after 6 months of treatment.

- Management of KHE depends on both lesion and patient characteristics, presence of coagulopathy, and physician preference.
- Topical tacrolimus has been reported to be effective for superficial KHEs and tufted angiomas in one small case series.[2] The exact mechanism of action is unknown.
- Spontaneous regression of KHE can also occur.
- Given the rapid improvement in this patient after initiation of topical treatment, we feel that resolution was achieved or accelerated here by his combination therapy.

Conclusions

- We report successful resolution of a superficial KHE with topical therapy alone.
- Topical treatment with tacrolimus and compression should be considered in select patients with superficial uncomplicated KHEs, thus avoiding potential complications associated with systemic treatment.

References:
1. Ji Y, Chen S, Yang K, Xia C, Li L. Kaposiform hemagioendothelioma: current knowledge and future perspectives. Orphanet J Rare Dis. 2020;15(1):39
2. Zhang X, Yang K, Chen S, Ji Y. Tacrolimus ointment for the treatment of superficial kaposiform hemangioendothelioma and tufted angioma. J Dermatol 2019;46(10):898-901