A rare case of kaposiform haemangioendothelioma involving the neck

Clinical History:

A 5-month-old female patient was brought in by mother for a rapidly growing right neck mass over 2 weeks. No fever or respiratory compromise. No known family history of malignancy or close contact with ill people. On examination, there was reddish discolouration on the mass. Laboratory investigations reviewed thrombocytopenia.

Imaging Findings:

Ultrasound (US) showed a solid subcutaneous neck mass with intralesional Doppler signals. On computed tomography (CT), the soft tissue mass is large and infiltrative with heterogenous enhancement. No calcification within. There is erosion and widening of the cervical neural foramina associated with intraspinal extension of the mass displacing the cord laterally. Right vertebral artery was encased but patent. Prevertebral and retropharyngeal extension noted as well causing mild airway narrowing. On magnetic resonance imaging (MRI), the mass appears isointense on T1, mildly hyperintense on T2 with moderate enhancement post-contrast. The intraspinal extradural extension was confirmed. No prominent tubular flow voids within. No cord oedema or leptomeningeal enhancement. No significantly enlarged cervical node. No intracranial extension. Overall findings are suggestive of kaposiform haemangioendothelioma (KHE) considering the clinical information of cutaneous discolouration with blood findings of Kasabach-Maritt phenomenon (KMP).

Following discussion with the paediatric team, US guided biopsy of the right neck mass was performed and histopathological assessment (HPE) revealed KHE. Child was referred to the paediatric oncology team for further management.

Discussion:

BACKGROUND

KHE is a rare vascular tumour of intermediate malignant potential derived from vascular endothelial cells. It commonly occurs in infants with usual involvement of the extremities, and rarely in the head and neck (HN) region as in our case. KMP is a laboratory blood findings highly associated with KHE, characterised by thrombocytopenic coagulopathy. Despite that, KMP can be absent as well in certain patients [1]. Distant metastasis is rare. There is no consensus yet on optimal treatment for HN KHE as for now, surgical resection and chemotherapy are usual treatments. Our patient was given Vincristine chemotherapy and steroids to control the KMP.
CLINICAL PERSPECTIVE

The typical clinical presentation is a rapidly growing subcutaneous mass with firm texture and ill-defined margins on palpation. Very often, there are overlying cutaneous reddish discoloration, or papules. Additional symptoms depend on the extent of local effects onto the trachea and oesophagus.

IMAGING PERSPECTIVE

The appearance on imaging can be non-specific and at times mimic the appearance of a sarcoma. MRI typically shows an infiltrative mass with ill-defined margins, involvement of multiple vascular planes with cutaneous thickening and stranding of the subcutaneous fat, haemosiderin deposits, less prominent superficial vessels, and destructive changes or remodeling of the adjacent bone [2]. Signal characteristics are mainly hyper- or isointense on T1WI, hyperintense or slightly hyperintense with speckled hypointense signals on T2WI with heterogenous enhancement post-contrast [3]. Such findings on its own often lead to a broad list of differential diagnoses, but when the clinical feature of cutaneous skin changes and KMP are present, KHE should be the diagnosis. Although associated destructive changes of the bones have been documented in few case reports [4,5], the intraspinal extradural extension of the mass was an unusual finding in our case.

OUTCOME

Prognosis is variable. A recent meta-analysis quoted a mortality rate of 7.4% among patients with HN KHE due to local effects and KMP, with recurrence rate of 35.8%. Surgical resection with clear margins is associated with the highest rate of cured patients [6].

TAKE HOME MESSAGE/TEACHING POINTS

An aggressive infiltrative looking neck mass on imaging in a patient with cutaneous skin changes and KMP is highly suggestive of KHE. Even in the absence of both clinical features, KHE should be always included in the differentials in the group of infants and early childhood.

Written informed consent for publication has been obtained.

**Differential Diagnosis List:** Kaposiform haemangioendothelioma of the neck with kasabach-meritt phenomenon, Rhabdomyosarcoma, Infantile fibrosarcoma, Congenital haemangioma

**Final Diagnosis:** Kaposiform haemangioendothelioma of the neck with kasabach-meritt phenomenon

**References:**


Wong, B. L. K et al. (2016). Kaposiform haemangioendothelioma of the head and neck. Critical Reviews in
Oncology/Hematology, 104, 156–168. (PMID: 27365122)
Description: Right neck soft tissue mass causing erosion of the C2 vertebral body around the right foramen transversarium. Origin: Department of Radiology, Sarawak General Hospital, Malaysia
Description: Widening of the right neural foramina with bony erosion from C1/C2 to C4/C5 levels.
Origin: Department of Radiology, Sarawak General Hospital, Malaysia
Description: The right neck mass has retropharyngeal extension (arrow), and is encasing the right vertebral artery (flow void is preserved) with intraspinal extradural extension (arrowhead). Spinal cord is normal. Note the infiltrative nature of the mass involving multiple soft tissue planes. Origin: Department of Radiology, Sarawak General Hospital, Malaysia
Figure 4

Description: The mass appears isointense on T1. Origin: Department of Radiology, Sarawak General Hospital, Malaysia
Description: Large infiltrative right neck soft tissue mass with intraspinal invasion indenting onto the spinal cord (arrow). Origin: Department of Radiology, Sarawak General Hospital, Malaysia
Description: Solid heterogeneously isoechoic mass with internal vascularity. Origin: Department of Radiology, Sarawak General Hospital, Malaysia