Kaposiform haemangioendothelioma: A radiological dilemma

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Abstract
Kaposiform haemangioendothelioma (KHE) is a rare, locally aggressive vascular tumour diagnosed in infancy or early childhood. KHE has a worse prognosis than infantile or congenital haemangioma due to its infiltrative nature and risk of developing Kasabach-Merritt Syndrome (KMS). This case report is about a 4-month-old girl who presented with progressive left neck swelling. She was diagnosed with KHE and thrombocytopenia suggestive of Kasabach-Merritt Syndrome (KMS). We describe the imaging appearance of KHE on ultrasound and magnetic resonance imaging. We also highlight the importance of integrating patient’s clinical history with the physical, laboratory and imaging findings for early diagnosis to prevent disease-related life-threatening complications.

Keywords: Kaposiform haemangioendothelioma, vascular neck lesion, Kasabach–Merritt, consumptive thrombocytopenia

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Introduction
Kaposiform haemangioendotheliomas (KHE) is a rare type of locally aggressive vascular tumour with a reported incidence between 0.07 and 0.091 per 100,000 children per year, typically diagnosed in infancy or early childhood [1]. KHE is associated with a poorer prognosis than infantile or congenital haemangioma, a disease usually found within the similar age group [1,2]. KHE with the presence of severe thrombocytopenia due to platelet trapping within the tumour, sometimes accompanied by haemolytic anaemia, and secondary coagulation abnormalities form the characteristic features of Kasabach-Merritt Syndrome (KMS); a life-threatening condition which according to Zhou J et al has incidence rate of 45.9% and resulting in high mortality rate of up to 30% [3,4]. Radiological knowledge of this rare tumour especially in ultrasound and magnetic resonance imaging (MRI) is crucial in reaching the diagnosis and, ultimately in guiding the appropriate management [5].

Case description
A 4-month-old girl presented with progressive left neck swelling for 3 months. It was first observed at day 40 of life as a small non-tender swelling under the chin that gradually enlarged and extended to the left side of the neck. No prior history of trauma or family history of malignancy. The swelling became discoloured from blackish to purplish over the interval of 1 month. Otherwise, the patient was active and exhibited no symptoms of airway compression, fever, or weight loss.

Physical examination revealed a large swelling over the left side of the neck measuring 13.0 x 8.0 cm with upper border at the submental region extending to posterior auricular region and the lower border at the supraclavicular region. The overlying skin appeared purplish to bluish, non-tender, firm in consistency with no bruit on auscultation (Figure 1). No other swelling or neck nodes were palpable. Flexible nasopharyngolaryngoscope revealed no significant findings other than slight fullness over the left lateral pharyngeal wall and dilated vessels at the posterior pharyngeal wall.

On admission, full blood count investigation revealed thrombocytopenia with platelet count of 14 x 10⁹/L, normal total white cell count (11.6 x 10⁹/L) and haemoglobin (9.8 g/dL). Full blood picture showed no abnormal mononuclear or blast cells, which did not
explain the thrombocytopenia; therefore, peripheral consumption or destruction must be considered.

The neck ultrasound (US) demonstrated an ill-defined heterogeneously hypoechoic solid mass at the left preauricular region extending to the left submandibular and neck regions measuring 4.5 x 6.3 x 5.3 cm (AP x W x CC). This lesion exhibited minimal low-flow internal vascularity on colour and pulse wave Doppler (Figure 2). No enlarged cervical lymph nodes.

In the subsequent MRI, the mass appeared heterogeneously isointense on T1-weighted image (T1WI), heterogeneously hyperintense on T2-weighted image (T2WI) and exhibited heterogeneous enhancement post-contrast. It measures about 4.7 cm x 4.7 x 4.4 cm (AP x W x CC) and was associated with extensive oedema involving underlying subcutaneous tissue, left neck muscles and the adjacent deep spaces (Figure 3). The mass was not suppressed on fat suppression sequence with no blooming artefact in the gradient echo sequences to suggest haemosiderin deposit or calcification. There was no serpiginous flow void to suggest arteriovenous malformation. Feeding vessels from the external carotid artery were identified on magnetic resonance angiography sequence (MRA) (images not shown). In view of the clinical suspicion of KMS based on the patient’s age, clinical and haematological findings, the features on MRI could suggest a vascular neoplasm such as KHE.

A trucut biopsy was carried out and revealed positive immunohistochemical and histological morphology results for CD31 & ERG, CD34, and Podoplanin which supported the diagnosis of KHE (Figure 4).

A combination of syrup sirolimus and syrup prednisolone was started according to her body weight as the first line of treatment. After 3 weeks on
Figure 3. Neck MRI in axial and sagittal T2WI showing heterogenous hyperintense mass (arrows) (a,c) which heterogenously enhanced on T1WI post-contrast obscuring multiple soft tissue planes (b,d).

Figure 4. Immunohistochemical and histological morphology results of left neck mass:
medication, the swelling on her left neck has reduced in size which clinically measured 7.0 cm x 6.0 cm (previously 13.0 x 6.0 cm) with minimal bluish discolouration. Syrup prednisolone was tapered off, while syrup sirolimus was continued with a plan to increase dose according to patient’s condition. After nearly 4 months on medication, the left neck swelling became much smaller with no bluish discolouration and softer on palpation (Figure 5).

Repeated MRI scan done at 10 months after the medication commencement showed significant reduction of the mass size which measures 2.7 cm x 1.8 cm x 2.3 cm (AP x W x CC) with residual lesion seen at the left submandibular space and below the left mandible (Figure 6). The patient remained active, developmental milestone was appropriate to age, and she had no other clinical symptoms.

Figure 5. Following 4 months of oral treatment, the left neck swelling has significantly reduced in size with no obvious skin discolouration.

Figure 6. Repeated neck MRI in axial (a) and sagittal (b) views showing the heterogenous hyperintense mass has significantly reduced in size with less locoregional tissue involvement (arrows).

a. Strip of tumour tissue composed of vague irregular lobules made up of proliferation of rounded capillary-type vessels (circle) and dilated lymphatic vessels (rectangular), embedded within oedematous, fibrous, and haemorrhagic stroma. Peripheral fibrosis and hyalinization of stroma is observed (HE stains 40x magnification).

b. ERG immunostaining (brown coloured stain) is positive and highlights the endothelial cells lined the spindled cells and vascular lumina.

c. CD31 positive highlights the endothelial cells. Some of the vascular lumina are filled with platelet-rich microthrombi, highlighted by CD31 immunostaining (red arrow).

d. Podoplanin (brown coloured stain) highlights the lymphatic endothelium.
Discussion

Vascular lesions within the head and neck have a broad pathological spectrum. The most used classification is from the International Society for the Study of Vascular Anomalies (ISSVA), which classifies the lesions into benign, locally aggressive/borderline, and malignant [1]. The actual prevalence and incidence of KHE are most likely higher than those reported, as smaller KHE lesions are typically not associated with KMS or other related conditions and can therefore be misdiagnosed as an uncommon variant of infantile haemangioma or other vascular anomalies [6].

To classify the lesion in this patient, radiological knowledge is required. Based on our ultrasound findings, a vascular lesion is suspected due to the ill-defined infiltrative pattern, and it exhibits low-flow colour and pulse wave on Doppler [7,8]. A well-defined lesion may be a haemangioma, while ill-defined lesions may represent KHE, angiosarcoma or another malignant lesion [7–9]. However, according to Bansal AG et al these lesions can be further differentiated by looking into the pattern of vascularity within the lesion which is divided into high-flow and low-flow [9]. In our case, the minimal intralesional vascularity with a slow-flow pattern on ultrasound favours towards lymphatic malformation, mixed venolymphatic or other common non-vascular tumour like rhabdomyosarcoma which can exhibit variable internal vascularity [9].

To further characterise the lesion, MRI is indispensable. In our patient, the mass appeared ill-defined, isointense to muscles on T1WI, and heterogeneously hyperintense on T2WI involving multiple planes of soft tissue with heterogeneous enhancement post-contrast, features which favoured KHE according to Brambhatt et al and Navarro et al [1,10]. Stranding of the surrounding subcutaneous fat, as observed in our patient, lends additional support to the diagnosis of KHE. However, other indicators of KHE, such as hemosiderin deposits and destructive changes of the adjacent bones, were absent in our patient [10]. In contrast, infantile haemangioma, despite having an incidence within the same age group, it is described as a well-defined solid homogenous mass that appears isointense to muscles on T1WI, hyperintense on T2WI involving multiple planes of soft tissue with homogenous enhancement post-contrast, features which favoured KHE according to Brambhatt et al and Navarro et al [1,10].

Conclusion

Correlation between patient’s history, clinical findings, laboratory results and imaging appearance is of paramount importance in diagnosing paediatric masses, especially when they are rare. MRI is an essential imaging tool to characterize vascular lesions. Early diagnosis is vital in preventing life-
threatening conditions that result from KMP or due to the nature of KHE.

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References