A neonate with Kaposiform haemangioendothelioma, complicated with Kasabach-Merritt phenomenon

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Introduction

Kaposiform Haemangioendothelioma (KHE) is a locally aggressive congenital vascular tumour arising from the vascular endothelial cell lining, frequently associated with the Kasabach-Merritt Phenomenon (KMP)[1]. Although KHE commonly occurs in the extremities, it can be observed in retroperitoneal and intrathoracic regions[1]. KHE has an incidence of 0.91/100,000 children, 60% manifesting neonatally[1]. Therapeutic options include pharmacotherapy, surgical resection, radiotherapy and embolisation[2,3]. We report a neonate presenting with KHE complicated by KMP.

Case report

A baby boy was delivered by elective caesarean section at 38 weeks of gestation, as antenatal scans revealed a mass in the left thigh. His birth weight was 2.8 kg, occipito-frontal circumference 34 cm and length 51cm. He had an erythematous swelling of the left thigh (Figure 1). There were no cutaneous haemangiomas elsewhere, and the baby was pink with no bleeding manifestations. Saturation of the left lower limb was 97% in room air.

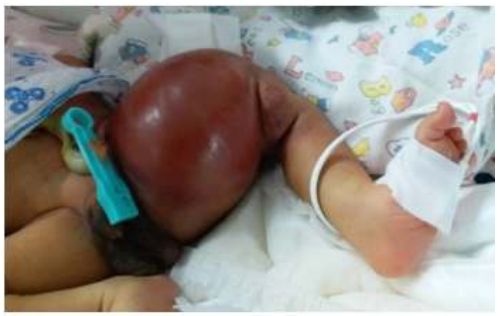

Figure 1: ***Erythematous swelling of the left thigh***

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On admission, the haemoglobin level was 11.2g/dL, the total white blood cell count was 9100/cu mm and the platelet count (PC) was 53,000/cu mm. Ultrasound scan of the lesion revealed a 11 x 10 x 8 cm in size, highly vascular, non-homogeneous, soft tissue area in the thigh, encircling the femoral artery. Magnetic Resonance Imaging (MRI) revealed an ill defined, large, soft tissue mass, predominantly subcutaneous, surrounding the thigh, extending from pelvis to the knee. Thigh muscles were enlarged with abnormal high signal in T2 with contrast enhancement. Femoral bundle was encased but normal in calibre. Femur was not involved (Figure 2).

Histological examination of the Tru-cut biopsy specimen revealed irregular infiltrating nodules of compressed vessels in a dense hyaline stroma with glomeruloid islands, characteristic of KHE (Figure 3). In addition, he developed severe thrombocytopenia (lowest PC 4,000/cu mm) and coagulopathy (activated partial thromboplastin time: 68 seconds, international normalised ratio 2.5) resulting in gastrointestinal and skin bleeding. Blood picture was compatible with microangiopathic haemolytic anaemia.

Due to involvement of multiple tissue planes, the lesion was not suitable for surgical resection. Therefore, medical management was instituted in the first week of life with oral dexamethasone and oral propranolol. The aim was to reduce the size of the lesion to prevent complications. Initial response to oral dexamethasone 1mg/kg/day and oral propranolol 2mg/kg/day was unsatisfactory. Therefore, weekly vincristine 0.025mg/kg was given for 6 weeks, under supervision of a paediatric oncologist. There was a marked response to vincristine, with reduction in the size of the lesion and improvement in haematological parameters. He was transfused with platelets as he was having bleeding manifestations with severe thrombocytopenia. The course was complicated with local infection at the biopsy site, for which the baby had to undergo incision and drainage several times, in addition to intravenous antibiotics. The baby showed a slow but complete resolution of the lesion at 4 months of age (Figure 4), along with resolution of the thrombocytopenia and coagulopathy.

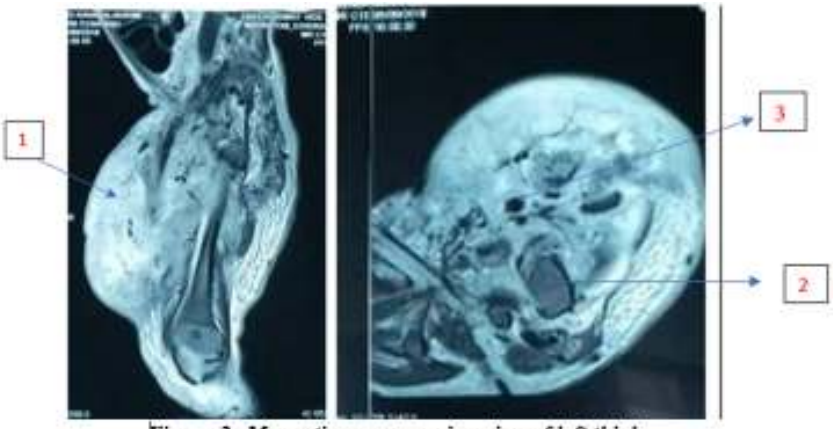

Figure 2: ***Magnetic resonance imaging of left thigh***

1. *Irregular soft tissue mass*
2. *Femur not involved, although encircled by the lesion*
3. *Femoral bundle is encased by the lesion*

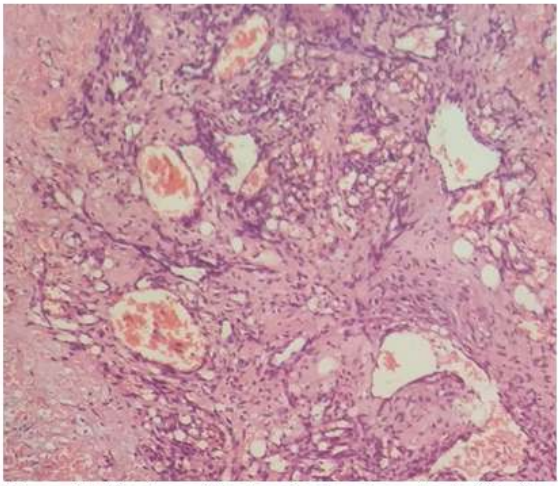

Figure 3: ***Histology of lesion showing irregular infiltrating nodules of compressed vessels in a dense hyaline stroma with glomeruloid islands***

Figure 4: ***Complete resolution of lesion***

Discussion

KMP is believed to arise from endothelial defects within the haemangioma, resulting in platelet activation leading to thrombosis formation, consumption of coagulation factors and elevated fibrinolysis[4]. Therefore, severe thrombocytopenia, hypofibrinogenaemia and raised fibrin split products are observed. Evidence of intravascular haemolysis is often present with fragmented red cells in blood film, high lactate dehydrogenase levels and hyperbilirubinaemia[4].

Contrary to popular belief that KMP can occur with haemangiomas, it has been elucidated that KMP occurs only with KHE and rarely with vascular neoplasms, and never with common haemangiomas[4]. The risk of development of KMP is higher in retroperitoneal and intrathoracic KHE, deep seated lesions and those involving muscles[1].

Despite severe thrombocytopenia, significant bleeding episodes are rare[5]. An important learning point is that platelet transfusions must be discouraged, unless there is active bleeding, or an invasive procedure is planned. Platelet transfusions can lead to aggravation of intralesional bleeding, increased platelet activation, facilitation of rapid tumour growth and exacerbation of pain[5]. Furthermore, the half-life of transfused platelets is very short[5]. Similarly, plasma products must be administered only if there is active bleeding or the child is being prepared for surgery[5].

Pharmacological therapy is one of the mainstays of treatment. Corticosteroids and propanol are used widely to treat KHE[1,2]. Second line treatment

includes vincristine and interferon alfa[3,5]. Despite toxicities (flu like symptoms and risk of spastic diplegia)[5], interferon alfa has been tried in some patients with favourable results and it has been particularly useful in steroid resistant lesions[5].

Surgical resection is useful for localized lesions. However, it is often not possible to resect totally, due to anatomical complexity[5]. Embolization is used for lesions with recognisable feeder vessels. Complications include ischaemic damage to organs, aggravation of haematological parameters and relapse[5].

Our patient responded to vincristine therapy. Vincristine therapy is associated with zero mortality[3]. According to literature, only 29% patients experienced complete resolution, while regression in size was observed in 43%[3].

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