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Management of a Kaposiform haemangioendothelioma of the kidney with Kasabach-Meritt phenomenon without chemotherapy

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ABSTRACT

Kaposiform haemangioendothelioma (KHE) is a benign vascular tumour. While it is not associated with distant spread, it can be locally aggressive and associated with Kasabach-Merritt syndrome (KMS). There were only 2 prior reports of KHE isolated to the kidney, with both having been managed with neo-adjuvant chemotherapy. We present a case report of renal KHE in a paediatric patient managed instead with upfront surgical resection, and discuss the close interdisciplinary diagnostic and management approach undertaken to facilitate this, and the considerations in the approach.

1. Introduction

Kaposiform hemangioendothelioma (KHE) is a rare vascular tumour typically seen during childhood. Most cases of KHE involve superficial and deep tissues of the extremities, while visceral involvement is less common. Kasabach-Merritt syndrome (KMS), characterized by profound thrombocytopenia and consumptive coagulopathy, is a potentially life-threatening complication commonly associated with KHE. We report a case of KHE occurring in the kidney in a 5 month old infant with associated KMS, and successful treatment of renal KHE with upfront surgical resection without neoadjuvant or adjuvant chemotherapy. This case highlights the importance of tight interdisciplinary coordination in the approach and successful management of a rare paediatric solid tumour.

2. Case report

A 5 month old girl presented to the outpatient general paediatric clinic with a 2 week history of an incidentally-detected left abdominal mass. She was antenatally well and born at 39 weeks' gestation with a birth weight of 3.36 kg. Postnatally, the child was in good health. There were no associated urinary symptoms or preceding weight loss. On physical examination, there was a ballotable mass over the left upper abdomen. Cardiac, respiratory and genital examinations were unremarkable. Her spine was normal with absence of scoliosis and neurological deficits. There was no hemihypertrophy or petechial rashes.

Computed tomography (CT) of the abdomen and pelvis revealed a 3.9 × 6.4 × 5 cm enhancing lobulated mass in the left retroperitoneum adjacent to the left renal hilum, displacing the left kidney inferiorly and invading the renal parenchyma at the lower pole. There were no internal calcifications or necrosis within the mass (Fig. 1). Renal Doppler ultrasound demonstrated marked left hydronephrosis due to extrinsic compression from the mass. Imaging was suspicious for an exophytic renal tumor and less likely a neuroblastic tumor encasing the renal hilar vessels.

Serum beta-hCG and alpha-fetoprotein, and urinary

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vanillylmandelic acid and homovanillic acid were unremarkable. Hemoglobin level was 8.2 g/dL (range: 12–16 g/dL), white blood cell count was 12,000/μL (range: 4500–13500/μL) and platelet count was 23,000/μL (range: 150,000–450,000/μL). This was of concern for possible bone marrow involvement by tumour. However, bone marrow aspiration and trephine biopsy showed trilineage marrow with erythoid and megakaryocytic hyperplasia with no evidence of tumour infiltration.

Haematology input was sought and differentials for peripheral causes of thrombocytopaenia included immune-mediated thromobocytopenia as a paraneoplastic phenomenon or thrombocytopaenia secondary to tumour sequestration. The patient was started on a trial of intravenous immunoglobulin and 3 doses of intravenous methylprednisolone. However there was no sustained response in platelet counts. Platelet trapping secondary to KMS was suspected, further evidenced by hypofibrinogenemia [fibrinogen 0.40 g/L (normal range 1.80–4.80 g/L)] and elevated markers of coagulation activation [D dimers >32 mg/L (normal range 0.19–0.55 mg/L). This raised the suspicion for a vascular anomaly as the underlying aetiology of the mass. In order to optimize preoperative platelet levels, a trial of transfusion with platelet concentrate was performed which showed a satisfactory rise in platelet count to a safe level for surgery, from 25,000/μL to 127,000/μL at 1 hour post transfusion, before decreasing to 22,000/μL by 24 hours post-transfusion. Thus, a left radical nephroureterectomy was performed under the cover of continuous platelet and cryoprecipitate transfusions (Fig. 2).

Intraoperative rotational thromboelastometry (ROTEM™) was used to guide the transfusion of blood products. Intraoperative INTEM and FIBTEM reflected the hypofibrinogenemia and a deficient intrinsic pathway. Intravenous tranexamic acid bolus and infusion was administered to minimize blood loss. Other strategies employed intraoperatively to minimize blood loss included mild hypotension and maintenance of normothermia via the use of active warming devices. The INTEM and FIBTEM repeated toward the end of surgery showed a robust clot formation with normal clot formation time, alpha angle, and maximal amplitude. The immediate postoperative haemoglobin was 10.9 g/dL, platelet count of 95×10^9/L and fibrinogen 4.44 g/L. The patient was transferred to the intensive care unit postoperatively and extubated the following day.

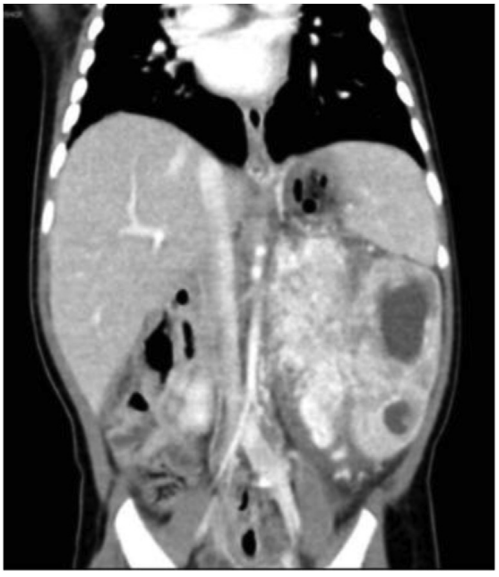

Fig. 1. Contrast enhanced CT scan demonstrating the exophytic lobulated mass centered at the left renal hilum, with prominent contrast enhancement and associated hydronephrosis.

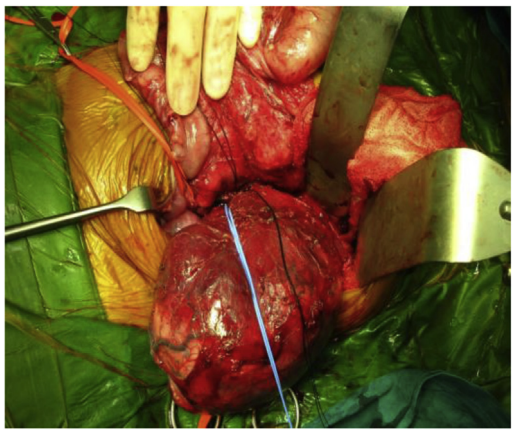

Fig. 2. Intraoperative view of highly vascular left retroperitoneal tumor contiguous with the medial aspect of the left kidney and adrenal gland, completely encasing the left renal hilar vessels isolated on blue and red vessel loops.

Her platelet trend increased to 292,000/μL by post-operative day 2, without further platelet transfusions. Her postoperative course was complicated by gastroparesis and ileus lasting 13 days, which was treated conservatively with intravenous erythromycin. In view of the histopathological findings, no adjuvant chemotherapy or interferon alpha was indicated. The patient returned to her home country and no complications were recorded in subsequent follow-up at 1 year after operation in our hospital.

Pathological examination of the nephroureterectomy specimen revealed a lobulated haemorrhagic tumour replacing the renal parenchyma with prominent hydronephrosis (Fig. 3a). Histology showed multiple blood-filled lobules of plump spindled tumour cells forming slit-like anastomosing and complex vascular spaces (Fig. 3b). The spindled tumour cells had oval nuclei containing dispersed chromatin and small nucleoli. Occasional fibrin thrombi were noted (Fig. 3c). Immunohistochemical stains showed positive reactivity for vascular markers CD31 and CD34. D2-40 was positive in patchy areas indicating lymphatic differentiation (Fig. 3d). There was negative staining for desmin, GLUT1, CD10 and WT-1. The histological features and immunophenotype were in keeping with

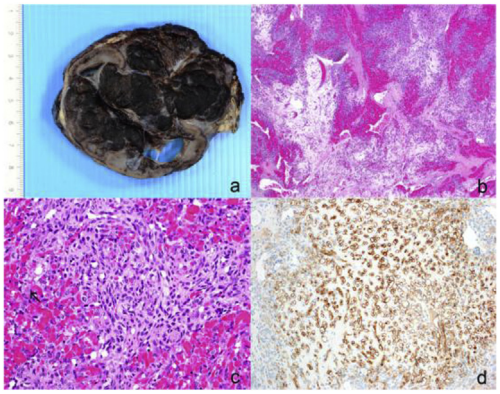

Fig. 3. a. Gross specimen showing lobulated hemorrhagic tumor replacing the renal parenchyma, **b.** Photomicrograph (H&E, × 20) demonstrating lobulated architecture of tumor and prominent congestion of vascular spaces, **c.** Photomicrograph (H&E, × 400) demonstrating complex well canalized to slit-like vascular structures lined by spindled cells with occasional fibrin thrombi (arrow), **d.** Immunohistochemical stain with D2-40 showing patchy reactivity indicating differentiation towards a lymphatic phenotype.