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Treatment of retroperitoneal kaposiform hemangioendothelioma: 2 case reports

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ABSTRACT

Kaposiform hemangioendothelioma (KHE) is an uncommon vascular tumor that affects young children. It frequently affects the trunk, limbs, head and face, but rarely the retroperitoneal area. This retrospective study analyzed two cases of retroperitoneal KHE admitted to the Children's Hospital of Fudan University, and investigated the clinical characteristics and treatment of this disease.

1. Materials and methods

Two patients with combined retroperitoneal KHE and Kasabach-Merritt phenomenon (KMP) were admitted to the Children's Hospital of Fudan University from 2009 to 2011.

2. Case 1

A five-month-old male child was admitted with a complaint of an "accidentally discovered abdominal mass noticed a week ago." Physical examination revealed a distended, left mid-to upper abdomen. Upon palpation, it was felt as a solid mass, 7 cm in diameter, with a clear boundary and no tenderness. Ultrasonic examination revealed a giant mass comprising a mixture of solid and liquid contents in the adrenal area of upper left retroperitoneum, indicating the possibility of neuroblastoma. The mass was located adjacent to the tailof pancreas and left kidney, resulting in the compression and deformation of the spleen. Abdominal enhanced CT scan revealed a huge mass with a clear boundary and measuring 7.4 × 7.7 × 10 cm in size, located in the left abdomen, exceeding the midline. The mass appeared as isodense to slightly hypodense lesions with stripe calcification. The lesions exhibited heterogeneous enhancement, while the spleen was not clearly visualized. Computed tomography angiography (angiography or CTA) showed that the blood supply of the mass originated from the branch of splenic artery (Fig. 1A). Complete blood count (CBC) revealed a platelet count of 27×10^9/L. Pre-operative diagnosis was "retroperitoneal mass with the possibility of neuroblastoma."

After platelet transfusions, exploratory laparotomy and mass biopsy were performed. During the operation, a giant solid tumor was found originating in the retroperitonium, which elevated the spleen and pancreas. Nodular projection was detected on the surface of the mass. The mass infiltrated the spleen, and could not be separated. Due to the unique pathological manifestation, no precise, short-term diagnosis was possible. After two weeks of consultation with the Pathological Department of Fudan University Shanghai Cancer Center, we finally diagnosed the tumor as (abdominal) Kaposiform hemangioendothelioma, based on biopsy.

3. Case 2

A four-month-old female child was diagnosed as volvulus based on "darkened feces accompanied with crying and vomiting" at a local hospital. During exploratory laparotomy, hemorrhagic enteritis was diagnosed and the child did not receive any special treatment. After postoperative hormone therapy and platelet transfusions, symptoms were relieved within a short period followed by recurrence. Physical examination revealed a pale appearance and a soft and distended abdomen. No tenderness or apparent solid mass was detected. The bowel sound was weak. CBC showed a hemoglobin value of 70 g/L; platelet count,

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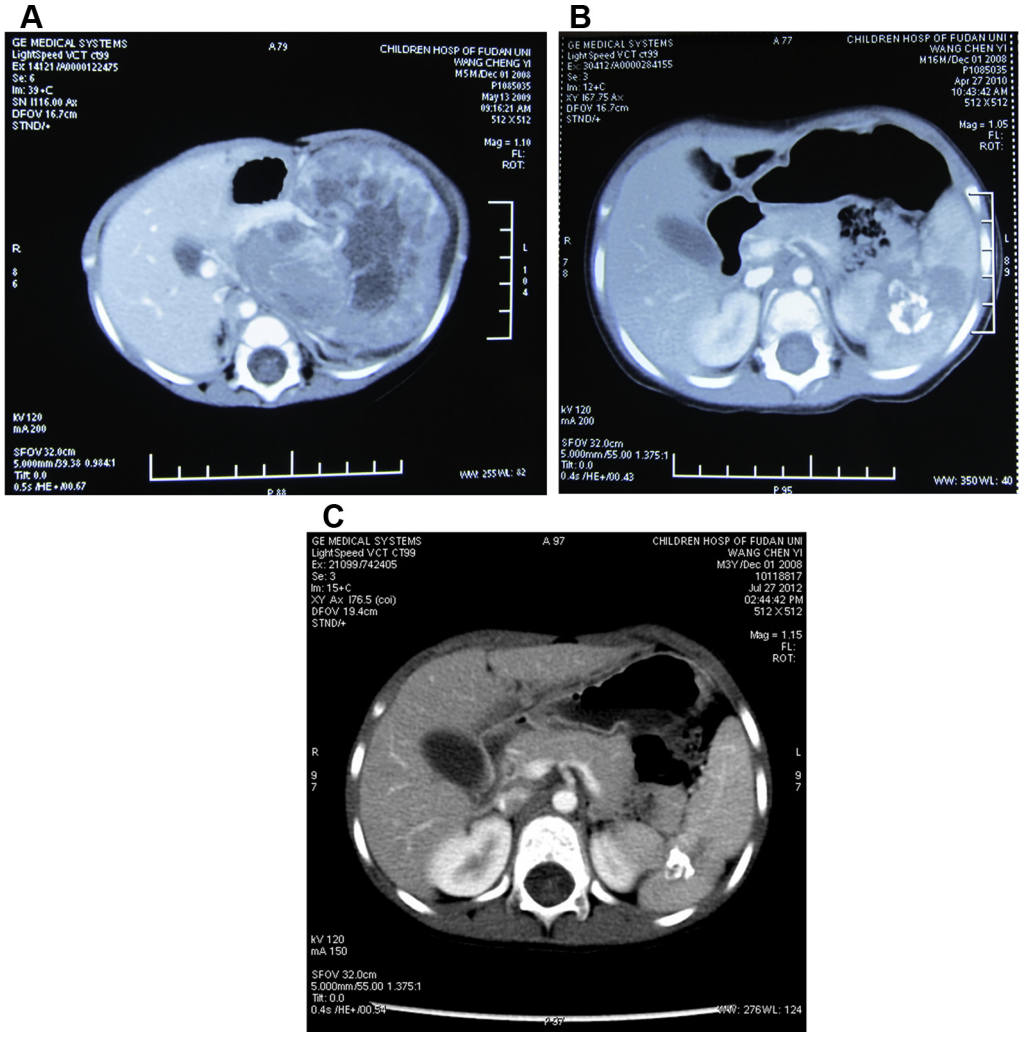

Fig. 1. A: Disease onset. A huge heterogenous mass with a clear boundary and a diameter of 7 cm, located in the left abdomen, exceeding the midline; stripe calcification can be seen. The lesions exhibited heterogeneous enhancement, while the spleen was not clearly visualized. B: One year after VCR treatment. The mass was diminished in size. Spleen was restored to normal position. C: Two years after VCR treatment. The tumor almost disappeared, with only slight calcification left.

12×10^9/L; and white blood count, 8.9×10^9/L. The child tested positive for fecal occult blood. Ultrasonic scan indicated abnormal bowel in the left abdomen. MRI revealed an abnormal crumby signal in the mesentery root. The intestine wall was thickened and swollen accompanied with a large amount of ascites (Fig. 2A). CTA revealed multiple abnormal enhancements in the head of pancreas and mesentery root. Intestine wall became thickened and swollen accompanied with a large amount of ascites and collateral vessel formation (Fig. 2B).

After consultation with multiple clinical departments, the child was diagnosed as retroperitoneal KHE combined with thrombocytopenia. Medical hemorrhagic diseases and strangulated intestinal obstruction were both excluded.

4. Results

4.1. Case 1

Due to the unique pathological manifestations, we finally certified the pathological results as retroperitoneal KHE after multidisciplinary consultation 2 weeks later. The child underwent experimental chemotherapy with cyclophosphamide (CTX), adriamycin and etoposide (VP16) on day 5, postoperatively. Persistent thrombocytopenia following chemotherapy and methylprednisolone pulse therapy was used to arrive at a definitive pathological diagnosis. After the treatment, platelet counts were rapidly elevated to 121×10^9/L. Oral drugs (5 mg/kg, qd) was then administered instead of intravenous methylprednisolone and platelet counts were maintained at 100×10^9/L. The tumor size was reduced by 25%, 1 month later. During the treatment phase, platelet counts decreased to 69×10^9/L prompting reduction of the dose of methylprednisolone and gradual substitution with vincristine (VCR) treatment comprising 10 cycles. The tumor almost disappeared although slight calcification was visible 2 years later (Fig. 1B and C).

4.2. Case 2

After consultation with multiple clinical departments, a definitive diagnosis of retroperitoneal KHE was obtained. The child received hormonal pulse therapy without any response. The child exhibited continuous bleeding and non-elevated platelet counts. VCR was used simultaneously and platelet counts were elevated immediately to 85×10^9/L, resulting in gradual hemostasis. After 4 cycles of VCR