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PHOTO CLINIC

Kaposiform Hemangioendothelioma of Abdominal Wall Associated with Kasabach-Merritt in A Newborn with Dramatic Response to Sirolimus and Vincristine

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A 26-day-old male baby was referred to the neonatal ICU from a rural area hospital with severe anemia and thrombocytopenia since birth. He was the second child of consanguineous parents. On physical examination, an extensive cutaneous purple to black lesion with ill-defined margins throughout the anterior abdominal wall musculature was visible which had caused some degrees of abdominal distension (Figure 1). The lesion was firm and indurated on palpation extended from subcostal area caudally to the iliac crest with a halo of ecchymoses extending up to around nipples bilaterally. Initial Laboratory tests showed Hb: 6.6 gr/dL, platelet: 7000/µL with normal white blood cell count. The baby was started on broad spectrum antibiotics and received multiple transfusions of FFP, packed RBCs and platelets. The coagulation profile revealed a prolonged prothrombin time with a very low fibrinogen (40 mg/dL), elevated fibrin degradative products (FDP) (more than 20 µg/mL by agglutination method) and D-dimer more than 10,000 ng/mL by ELFA method (Normal<500 ng/mL). Accordingly, a diagnosis of Kasabach-Merritt syndrome was made for the newborn in view of severe thrombocytopenia, anemia, severe coagulopathy, and the huge abdominal wall lesion which was in favor of a vascular tumor.

Oral prednisolone (2 mg/kg/day) and propranolol (1 mg/kg/day) was started; however, no improvement was seen in the size of the lesion or hematological abnormalities. A skin punch biopsy of the lesion was performed on sixth week of life which was reported as "kaposiform Hemangioendothelioma" (Figure 2). Spiral CT angiography of abdomen showed a large



Figure 1: The extensive cutaneous purple to black lesion with ill-defined margins throughout the anterior abdominal wall musculature with invasion to subcutaneous tissues.

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Figure 2: The vascular lesion of the abdominal wall after skin punch biopsy.

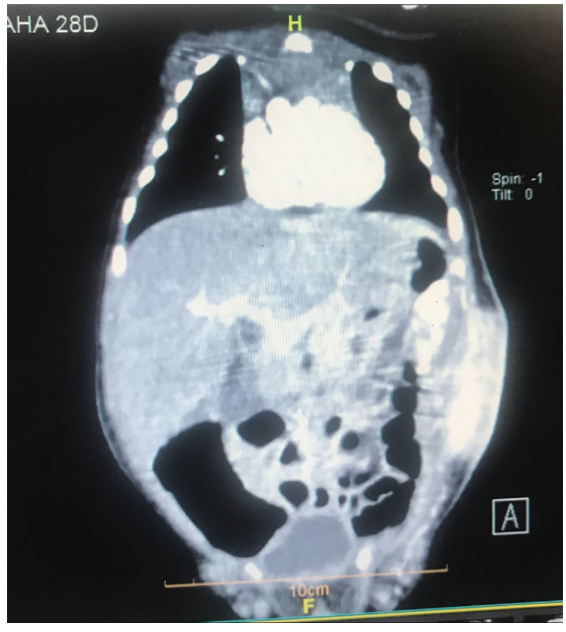


Figure 3: Spiral CT angiography of abdomen showed a large enhancing mass in left side of the abdominal wall involving left oblique and transverse abdominis muscles along with transversalis fascia in medial aspect.



Figure 4: Significant regression of the cutaneous KHE after four weeks of treatment with vincristine and sirolimus.



Figure 5: There was complete regression of the KHE without any induration with only discoloration of the skin on follow up after one year.

enhancing mass in left side of the abdominal wall involving left oblique and transverse abdominis muscles along with transversalis fascia in medial aspect (Figure 3). Superior and inferior epigastric arteries in left side were enlarged and tortuous supplying the mentioned lesion. The abdominal aorta was normal. The treatment protocol was changed to oral sirolimus (0.8 mg/m^2/dose, twice a day) and weekly injections of vincristine in addition to propranolol while the steroid was tapered off over 2 weeks.

The patient responded dramatically to sirolimus and vincristine with hematologic recovery and complete normalization of platelets in two weeks. Upon repeated CT scan, a significant reduction in the size of the abdominal wall lesion in four weeks was observed. The cutaneous lesion was also regressed clinically (Figure 4). The treatment was continued with weekly vincristine for 6 weeks and then every two weeks to monthly intervals and oral sirolimus for one year. The lesion of the abdominal wall was replaced by subcutaneous fat without enhancement on last abdominal CT scan. There was no lesion or induration palpated clinically with only discoloration of the skin on follow up after one year (Figure 5).

KHE was first described in 1993 by Zukerberg and colleagues as a distinctive lesion of infancy and early childhood with common features to both hemangiomas and Kaposi's sarcoma. It is a rare vascular tumor typically seen as a cutaneous lesion with ill-defined borders.[1] There is still a paucity of literature regarding the incidence of KHE. According to the recent World Health Organization (WHO) classification of soft tissue tumors, KHE belongs to the subgroup of rare vascular tumors with intermediate malignant potential which are locally aggressive with a high rate of local recurrence.[2] In the largest series of KHE, Croteau et al. from Boston Children's Hospital have registered 107 children from 15 countries during an 18-year-period. The prevalence of KHE is reported

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about 0.9 case per 100 000 children with 93% of cases in infancy and 60% as neonates. Eighty-nine percent of the patients had a cutaneous lesion, while no patient had multifocal lesions. Cutaneous discoloration and progressive enlargement of the tumor occurred in 75% of cases.[3] Our case demonstrated a very large cutaneous lesion with dark blue discoloration overlying almost the whole abdominal wall, which was firm, thickened and indurated on palpation. Cutaneous KHE more commonly involved the extremities, especially overlying the joints. However, in the cohort from Boston Children's hospital, 11% of patients lacked any cutaneous involvement.[3] KHE is an infiltrative tumor that may cross tissue planes from dermis into subcutis, fascia, muscle, and the bone.[4] In another cohort study of 146 patients diagnosed with KHE, the median age at diagnosis was 2·3 months and the extremities were involved in 50.7%. Cutaneous lesions with deep infiltration comprised 63% of the lesions.[5] The lesion of our case also showed evidence of infiltration of the tumor to the underlying fascia and abdominal muscles on CT angiography.

Another characteristic aspect of KHE is its association with Kasabach-Meritt phenomenon (KMP) which was the presenting manifestation in our case. KMP is defined as profound thrombocytopenia associated with consumptive coagulopathy and hypofibrinogenemia which is defined to be observed only with KHE and tufted angioma (TA).[6, 7] KMP is reported to occur with an estimated incidence of 42 to 71%.[3, 5, 8] In the study from China on 146 children with KHE, 70% of patients developed KMP. Patients with KMP were more likely to have major complications. Young age (<6 months), trunk location and large lesions (>5 cm) were more complicated with KMP.[5] Abdominal wall involvement in KHE is very rare with 6 (chest or abdominal wall) out of 22 patients, ranging in age from 13 days to 7 years has been described.[9] In a report from Korea, twelve cases of KHE have been recognized in children with median age of 3 months (age range 7days-18 years) which only one case showed abdominal wall involvement.[10] There are few reports of multifocal involvement in KHE. A 13-year-old boy with KHE and multiple involvement in left elbow with extension to the arm and upper forearm, spleen, bilateral pulmonary masses with pleural effusion and multiple bone involvement has been described as a unique case of multifocal KHE.[11]

We started sirolimus besides the conventional treatments of prednisone and vincristine for the infant which resulted in a dramatic response. It is recommended as a first-line therapy or as part of a multidisciplinary approach for the treatment of KHE, specially in patients with KMP/KHE.[12-15]

Conflict of Interest: None declared.

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