Pancreatic Kaposiform Hemangioendothelioma Presenting with Duodenal Obstruction and Kasabach–Merritt Phenomenon: A Neonate Cured by Whipple Operation

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Abstract

Aim Kaposiform hemangioendothelioma (KHE) is a rare vascular tumor, commonly associated with Kasabach–Merritt phenomenon characterized by thrombocytopenia and consumptive coagulopathy. We report a case of pancreatic KHE presenting with neonatal duodenal obstruction and Kasabach–Merritt phenomenon.

Case Report A full term male baby presented with bile stained vomiting on Day 3 of life. Contrast study and computed tomography scan showed duodenal obstruction by a 5 cm extrinsic hypervascular mass. Platelet count was 23 x 109/L. Laparotomy confirmed a vascular tumor arising from the pancreatic head compressing on the duodenum. Whipple operation was performed.

Results Intestinal obstruction and thrombocytopenia resolved after surgery. There was no post-operative complications. Histology confirmed KHE. The boy was tolerating hydrolyzed milk formula and was thriving at 5 months follow up.

Conclusion We reported a case of pancreatic KHE presented with neonatal intestinal obstruction and Kasabach–Merritt phenomenon. High index of suspicion is necessary for diagnosis. To our knowledge, this is the youngest patient who underwent Whipple operation.

Introduction

Kaposiform hemangioendothelioma (KHE) was first described by Zukerberg et al in 1993.¹ It is a rare vascular tumor with an estimated prevalence of less than 1 in 100,000 children. It is more commonly seen in neonates and early infancy. In approximately 70% of KHE, Kasabach–Merritt phenomenon (KMP) occurs characterized by consumptive coagulopathy and thrombocytopenia.² KMP is rarely seen in the majority of common vascular tumors such as infantilehemangioma.³ KHE usually has a cutaneous origin affecting extremities, cervicofacial and torso body wall.⁴ Being classified as a vascular neoplasm, it usually infiltrates deeper tissue.⁵ KHE with visceral origin is exceedingly rare. We hereby report a neonate with KHE originating from the head of pancreas.

Case Report

A Chinese male baby with uneventful antenatal history except maternal gestational diabetes was born at 41 weeks by emergency cesarean section because of cephalopelvic disproportion. The birth weight was 4.18 kg. Apgar score was 8 at 1 and 5 minutes. Because of maternal gestational diabetes, he had blood investigation for complete blood count, plasma electrolytes, and glucose. He was transferred to the neonatal unit for incidental finding of thrombocytopenia with platelet
Clotting profile was normal and there was no clinical evidence of bleeding tendency. White cell count was 24.8 \times 10^9/L with C-reactive protein (CRP) of 13 mg/L. He was initially managed as perinatal sepsis with intravenous penicillin and gentamicin given. He developed bile-stained vomiting on day 3. Contrast study showed anterior and lateral displacement of the first and second part of duodenum with appearance suggestive of stretching by an extrinsic mass (Fig. 1). Abdominal sonography was performed on next day showing a retroduodenal hyperechoic mass measuring 3.8 \times 3.4 \times 5.2 \text{ cm} with increased Doppler signals. Computed tomography confirmed a retroperitoneal hypervascular mass close to the uncinate process of pancreas (Fig. 2).

Because of the persistent thrombocytopenia (platelet count of 37 \times 10^9/L) and duodenal obstruction, laparotomy was performed on day 7 revealing a 4 \times 4 \times 5 \text{ cm} retroperitoneal hypervascular tumor encasing the pancreatic head and lower end of common bile duct (CBD), displacing the duodenal C-loop, and causing extrinsic compression. The mass had feeding vessels from the gastroduodenal artery and drained to the superior mesenteric vein (SMV). Frozen section showed a vascular tumor with no evidence of malignancy. Attempted resection of tumor preserving the pancreatic head and CBD failed because of its hypervascular and infiltrative nature. Whipple operation was then decided with removal of the gall bladder, CBD, pancreatic head, distal one-third of stomach, pylorus, duodenum, and 15 cm of proximal jejunum with the tumor en bloc safeguarding hepatic artery, SMV, and portal vein. End-to-end pancreaticojejunostomy with dunking anastomosis (Fig. 3), hepaticojejunostomy, and gastrojejunostomy were performed. Estimated intraoperative blood loss was 393 mL. Postoperatively, the baby recovered gradually without complications. Platelet count returned to 257 \times 10^9/L. Oral feeding was started on day 10 and full feeding with normal formula was attained on day 14. Serum amylase and glucose was normal. He was discharged home 17 days after surgery.

The resection specimen was sent for histological examination. The pancreas was almost completely replaced by a tumor with closely packed lobules composing of proliferated oval and spindly endothelial cells forming whorls of Kaposi sarcoma-like sweeping fascicles, compact capillaries, and glomeruloid structures. On immunohistochemical examination, the neoplastic endothelial cells expressed...
Pancreatic KHE with Duodenal Obstruction and KMP  Leung et al. 9

ERG and CD31 but were negative for glucose transporter 1 and human herpesvirus 8. Pathological diagnosis was KHE.

The baby was followed up regularly after the operation. At 3 months, he developed steatorrhea and had failure to thrive with body weight fallen at third percentile. Serum albumin was 23 g/L although random glucose remained normal. Clinically, pancreatic insufficiency was suspected and high caloric, hydrolyzed formula feeding was prescribed. Body weight gradually improved to the 10th percentile at 5 months. Serum albumin increased to 37 g/L. He has been regularly followed up in the multidisciplinary nutrition clinic.

Discussion

KHE is classified as a vascular neoplasm according to the International Society for the Study of Vascular Anomalies (ISSVA). It is a rare high flow vascular tumor commonly affecting infants. It is locally aggressive and infiltrates surrounding tissues. To date, less than 300 cases have been reported worldwide. It shares the common histological feature of hemangioma and Kaposi sarcoma. Most KHE have cutaneous origin from extremities, torso body wall and cervicofacial regions. Retroperitoneal KHE is very rare with no more than 30 cases reported in the English literature. From the Boston series, approximately 12% of KHE originated from retroperitoneum. In 2009, we reported a retroperitoneal KHE infiltrating the mesentery causing intestinal obstruction and KMP. In approximately 70% of KHE, thrombocytopenia occurs as a result of platelet aggregation induced by turbulence blood flow within the distinctive endothelial architecture, leading to consumptive coagulopathy. It is not yet known whether KMP is associated with the site and size of KHE. In general, KHE with KMP is an aggressive lesion with poor prognosis. The platelet count will return to normal only after excision of the KHE. Platelet transfusion can cause painful enlargement of lesion and should be avoided without definitive treatment.

Pancreatic tumor is exceedingly rare in neonates and infants. Differential diagnosis of pediatric pancreatic tumor includes pseudopapillary tumor, pancreaticoblastoma, rhabdomyosarcoma, endocrine cell carcinoma, and vascular tumor. Muller et al described 18 pancreatic tumors in a 22-year period, with 11 pancreaticoduodenectomy performed. Vogel et al reported three pancreatic vascular tumors (two infantile hemangiomas and one KHE) in a 10-year database of over 5,000 vascular anomalies. Yeap et al reported three children between 1.5 and 8 years with pancreaticoduodenectomy performed over a 9-year period. Our patient had Whipple operation performed on day 7 of life with a body weight of less than 4 kg. To our knowledge, this is the youngest reported patient with successful Whipple operation in the English literature. The procedure is quite similar to that described in adult patients. However, pancreatocojunostomy is a major challenge. Dunking anastomosis is performed, requiring mobilization of the distal pancreatic stump and its invagination into the jejunum.

There are very few reports in the literatures describing the nutritional and endocrine status of children after pancreatic resections. Sugito et al followed up six patients after pancreatic resections. Two of them had increased hemoglobin A1c. From the French review of 18 pediatric pancreatic resections over a median follow-up period of 4.2 years, all patients had normal growth. There was no diabetes mellitus, but one patient had fat intolerance requiring pancreatic enzyme substitution. The postoperative plasma glucose level in our patient is normal. However, he developed indolent malabsorption while on normal infant formula with subsequent steatorrhea and failure to thrive. After implementation of hydrolyzed milk formula with increased medium chain triglyceride, the steatorrhea resolved and weight gain improved. We believe that long-term follow-up of this patient is essential to monitor his nutritional and endocrine status.

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