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Hepatoblastoma Related Tumor Thrombosis Extended to IVC, Ra and Pulmonary Artery Branches: A Case Report

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Abstract

Hepatoblastoma (HB) is a rare malignant tumor of the liver; but is the most common malignancy of liver in infancy and childhood that usually occurs in the first three years of life. These tumors are usually present by abdominal mass, abdominal pain, anemia, thrombocytosis, and high serum levels of AFP; and may cause decreased appetite and weight loss.

Complications such as ascites, GIB, icterus, splenomegaly and vascular thrombosis may occur, while hepatic vein or inferior vena cava thrombosis is quite rare especially with a spread to right atrium and pulmonary artery branches. Multidisciplinary therapy by a pediatric oncologist, a pathologist, an experienced radiologist, a pediatric cardiologist and a pediatric surgeon is very important to diagnose and manage the condition. We report on the case of an infant with a huge abdominal mass and much extended tumor thrombosis till the pulmonary artery branches that shrunk appropriately after the first course of chemotherapy.

Key Words: Abdominal mass, hepatic malignancy, Hepatoblastoma, Malignant liver tumor, Tumor thrombosis.

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1- INTRODUCTION

Hepatoblastoma (HB) is a rare malignant tumor of liver but is the most common malignancy of liver in infancy and childhood and includes ⅔ of pediatric hepatic malignancies and usually presents as abdominal mass. It has metastasized in more than ⅔ of cases at diagnosis. Some complications such as ascites, GIB, icterus or hepatosplenomegaly may more probably occur but hepatic vein or inferior vena cava thrombosis is quite rare especially with a spread to right atrium and pulmonary artery branches; and there are just few case studies reporting the coexistence of this rare phenomena and Hepatoblastoma.

2- CASE REPORT

Our case is a 22-month-old girl who was presented with abdominal pain from a month ago and abdominal enlargement from 2 weeks ago. A growing inelastic mass was palpable for the last 10 days in the right flank, RUQ, epigastric region and LUQ (liver edge 8cm subcostally). The patient was ill, tachypneic (RR= 69) with intercostal; and had low appetite, fever, postprandial vomiting and left lower limb edema.

Laboratory assays, including βhCG and AFP, abdominal sonography and CXR were done. Abdominal sonography revealed a heterogeneous solid mass as large as 118 ×69 mm in diameter that had filled up the left liver lobe entirely. Core needle biopsy of hepatic mass revealed pure epithelial (mixed fetal and embryonic type) hepatoblastoma. More evaluations with thoraco-abdomino-pelvic CT scan showed a filling defect in IVC extended up to right atrium suggestive of tumor thrombosis and right pulmonary artery and lobar branches suggestive of thromboembolie. (**Fig. 1**).

Moreover, multiple hypodense signals were seen in pulmonary parenchyma bilaterally as pulmonary metastases.

In echocardiography, normal systolic functions with a mass from IVC to RA (29×14mm) and two other masses on the free wall were found (11×8mm and 7×15mm). Coronary arteries were normal.

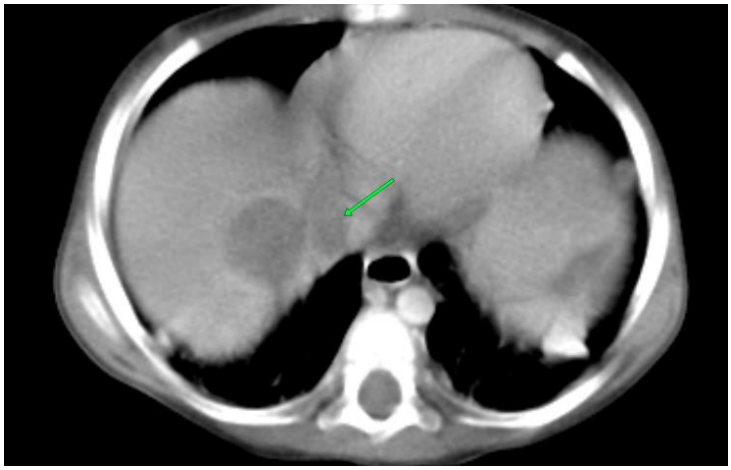
Previous medical data showed a thrombocytosis (plt=912000) with normal WBC and Hb indicating the likelihood of the relationship between this thrombocytosis and the hepatoblastoma diagnosed 7 months later.

After the first course of induction chemotherapy with cisplatin and doxorubicin, a desirable response was seen and tumor size and even tumor thrombosis of RA had been shrunk. Regardless of the admissible improvement of hepatoblastoma, nearly 18 days after the start of the treatment, the respiratory and mental state deteriorated again and finally we lost this sick child.

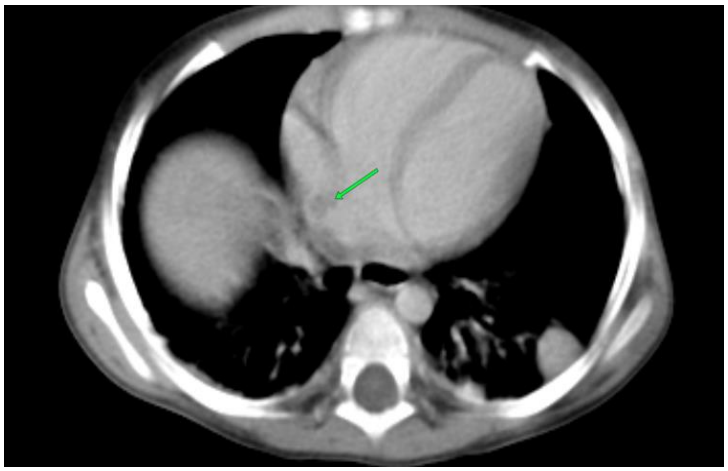
3- DISCUSSION

Around 120 years ago, the first case report of hepatoblastoma was published as “A Case of Tratoma Hepatis” in 18981 (1, 2). Hepatoblastoma is the most common pediatric malignancy of the liver (⅔ of cases) and usually presents by asymptomatic mass in the right and upper portion of the abdomen accidentally and more than ⅔ of these patients have metastases at diagnosis. Other problems such as anorexia, pain, weakness and weight loss may also occur (2, 3). Some complications would also happen such as ascites, GIB, icterus and splenomegaly. One of the rare complications of hepatic malignancies is thrombosis in the hepatic vein and IVC that is less prevalent in HB versus HCC (3, 4).

Thrombosis in the hepatic vein and IVC is almost rare but extension to RA and especially to pulmonary branches is quite rare and there are just few related case reports.



(a)



(b)

Fig. 1: The green arrow shows tumor thrombosis in suprahepatic IVC (a) and right atrium (b).

Loh et al. explained two cases of hepatoblastoma that both showed adequate responses in regard to the tumor size and tumor thrombosis spread following chemotherapy and thrombosis shrink upto below the diaphragm (5). Our case improved clinically in respiratory rate, vomiting, abdominal distention and tumor size (>50% declined), but no change was observed in the length of the tumor thrombosis and O2 saturation.

Another patient was described in a case study by Sarper et al. His advanced HB had remained a blue moon in remission just with surgery and no chemotherapy (6). Regardless of the fair response of our case to chemotherapy, she died and we lost the chance of more follow up.

The incidence of pulmonary metastases in HB with extended tumor thrombosis, IVC and RA is 100% (7). Our patient had multiple pulmonary metastases at diagnosis.

Endo et al. reported on a 4-year-old girl with HB (diagnosed according to open biopsy) in 2016 that her tumor thrombosis had extended up to RA. The tumor had responded desirably to 7 courses of neoadjuvant therapy but thrombosis had remained in RA and was, finally, evicted by extracorporeal circulation (8).

Hu et al., in 2020, illustrated that the most common sites of metastasis are blood vessels, brain and extrahepatic abdominal organs; moreover, the lungs are the most common sites of distant metastasis; and, finally, the existence of distant metastasis at diagnosis is the most important factor of poor prognosis (9). Indeed they didn′t explain the prevalence of the tumor thrombosis in their 36 patients, albeit their statement about the vascular involvement might have been indicative of intravascular tumor thrombosis. Our patient had no brain or extrahepatic abdominopelvic organ metastasis (according to MRI).

Faneli et al., in 2021, studied pediatric tumors with vascular involvement, comprising the renal tumors, adrenal tumors and hepatoblastoma. In this article, a patient with HB was treated by resection of tumor thrombosis of IVC and RA; and afterwards the resection of hepatic tumor. Moreover, the patient tolerated hepatic transplantation but died because of transplantation side effects. The researchers recommended that the optimal treatment approach for these patients requires an accurate teamwork by pediatric oncologists, pediatric radiologists, pediatric surgeons, heart surgeons and pathologists (10). Our case was not an appropriate candidate for surgery, according to her large tumor mass and respiratory distress; so, the only remained treatment option was chemotherapy, albeit after the initial favorable response to the first neoadjuvant therapy, she suffered from mental problem and respiratory deterioration and eventually died.

4- CONCLUSION

In this report, we aimed to discuss severe and extended vascular involvement with tumoral thrombosis in HB as a rare complication. Our assertion is that its optimal management requires the coherent and accurate cooperation between pediatric oncologists, pediatric cardiologists, pediatric surgeons, vascular surgeons, heart surgeons, radiologists (practiced in pediatric oncology) and experienced pathologists.

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