

Infantile Choriocarcinoma of the Liver: Case Report and Review of the Literature

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Abstract

We report our experience with a 3-month infant who presented with abdominal distension due to a huge hepatic mass and heart failure. Contrast enhanced CT scan was performed and showed a large hepatic mass with peripheral enhancement corresponding to infantile choriocarcinoma of the liver based on clinical, laboratory and imaging findings.

Keywords: Choriocarcinoma; Infantile; Liver

Introduction

To our knowledge, there are 32 cases in the English literature and all the published 32 cases are case reports [1-8]. Infantile choriocarcinoma of the liver is an extremely rare tumor originating in the placenta during gestation and detected in the perinatal and early infancy period as a hepatic mass. The reported outcome has been fatal in most of the published cases with a high tendency for metastasis. From an Imaging and clinical prospective, the radiologic appearance of hepatic choriocarcinoma is very like other pediatric liver tumors especially hemangioendothelioma and hemangioma. The description of the radiological features is rather scarce in the literature.

In this case report, we describe a 3-month-old baby boy who presented with a huge abdominal mass, failure to thrive and anemia. The aim of this paper is to familiarize the reader with the radiologic features, differential diagnosis, and management pathways of this entity.

Case Report

A 3-month-old male patient presented to the emergency department with pallor, shallow breathing, lethargy, generalized edema, and history of failure to thrive. The patient was hypotensive with blood pressure of 80/30 mmHg, tachycardia with heart rate of 170 beats/minute. On examination, the patient had abdominal distention with abdominal girth of around 54 cm. There are bruises in the abdomen with prominent veins. Hepatomegaly was palpated with abdominal mass noted within. The patient had low hemoglobin level of 5.9 g/dL (9.5 – 14 g/dL) and hematocrit of 20% (28% to 42%), PH of 7, PCO₂ of 110 and bicarbonate of 20 mmol/L.

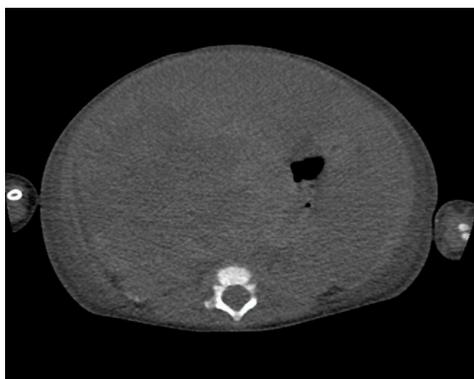


Figure 1a: Non-enhanced CT scan of the abdomen at the level of the liver show large hepatic mass with peripheral hyper-density and central hypo-density. No associated calcifications or fat components.

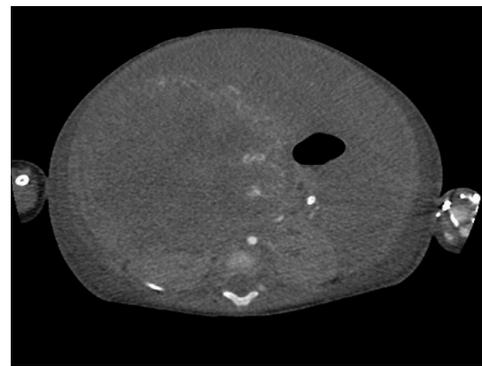


Figure 1b: Enhanced CT scan of the abdomen in the arterial phase with peripheral nodular pattern of enhancement similar to the enhancement of the aorta.

The patient was in severe respiratory distress and Critical Care Response Team (CCRT) was informed and the patient was shifted to the pediatric intensive care unit immediately. The patient was intubated and shortly required high frequency ventilation due to hypoxia and acidosis. He required multiple blood, plasma and platelets transfusions during his hospital stay. Due to associated coagulopathy, the patient received cryoprecipitate, vitamin K, tranexamic acid, and factor VII transfusions.

The history, lab and examination findings raised the concern of intraperitoneal bleeding presumably due to a hepatic mass. Therefore, an urgent CT scan of the abdomen was done. The obtained Contrast-enhanced CT scan of the abdomen and pelvis revealed a large heterogeneous well defined hepatic mass occupying the right liver lobe and showing peripheral hyper-density and enhancement post contrast administration with gradual filling and central necrosis (Figures 1a-1d). No calcifications, fat or cystic changes were associated. The adjacent liver parenchyma showed Nutmeg appearance due to

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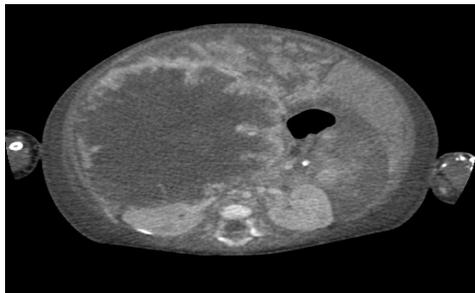


Figure 1c: Enhanced CT scan of the abdomen in the venous phase show increased filling of contrast in the hepatic mass with central necrosis. Note the enhancement of the mass continue to be like the aorta. The adjacent hepatic parenchyma show Nutmeg appearance due to hepatic congestion (*).

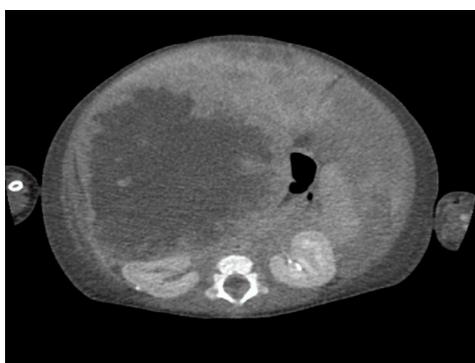


Figure 1d: Enhanced CT scan of the abdomen in the delayed phase show continued enchancement of the mass with central necrosis.

hepatic congestion. There was a peripheral wedge-shaped hypo-density within the spleen in keeping with splenic infarct. There was also associated small amount of hyper-dense free fluid likely representing blood. The lower cuts obtained through the chest were notable for a small pericardial effusion. Fluid overload was also noted in the form of diffuse anasarca. Based on these findings a preliminary diagnosis of hemangioendothelioma was given with the differential diagnosis of hepatoblastoma and mesenchymal hamartoma. At the time, infantile choriocarcinoma was not included in the differential mainly due to the rarity of the entity and the lack of specific findings to suggest it.

A panel of serum tumor markers (beta-human chorionic gonadotropin [B-HCG] and alpha-fetoprotein was obtained, revealing a markedly elevated corrected serum β -HCG level of 1125000 mIU/ml. The alpha-fetoprotein, which can be elevated in hepatoblastoma was within normal limits for age, measuring 8.4 ng/ml. The markedly elevated β -human chorionic gonadotropin suggested choriocarcinoma as the diagnosis. The presumptive diagnosis was made based on the clinical, imaging and tumor marker findings. A biopsy was performed during laparotomy to alleviate increased intra-abdominal pressure and unfortunately was insufficient; however, it showed necrotic tumor.

The patient continued to have refractory hypoxia and metabolic acidosis. He was started on chemotherapy on day 2 of admission. His septic search showed candida albican in respiratory and blood cultures. Unfortunately, he passed away 19 days after admission due to multi-organ failure despite of supportive measures. Of note, the infant's mother was choriocarcinoma free over five months' post-partum.

Discussion

Preclinical Following neuroblastoma and Wilms tumor, the liver

is the most common site for abdominal malignancy in children. The most common neoplasm involving the liver in children is metastasis most common primary liver neoplasms in children are malignant, about one-third are benign and may be of mesenchymal or epithelial origin [9].

Traditionally the differential diagnosis of hepatic tumors in children is based on age where hepatoblastoma, infantile hemangioendothelioma, and mesenchymal hamartomas are seen in the 0-5-year age group. Above 5 years the most common tumors are hepatocellular carcinoma and undifferentiated embryonal sarcoma. Metastatic lesions may occur at any age. Both hemangioma and infantile hemangioendothelioma may occur concurrently.

Most children with hepatic tumors commonly present with abdominal discomfort, palpable mass, and abdominal distension. Fetal and neonatal presentations include hydramnios, hydrops, heart failure, and respiratory distress. Occasionally, affected children may present with vomiting, fever, and clinical signs of abdominal irritation, suggestive of tumor rupture. To achieve a diagnosis, it is essential to combine age, gender, clinical and laboratory findings with radiologic appearance. Usually the findings on imaging either by US, CT or MRI are not only enough to reach a specific diagnosis but also guide the management of this subset of patients [10].

Infantile choriocarcinoma is a highly malignant sub-type of germ cell tumor and is thought to originate from the placenta as a complication of choriocarcinoma of the placenta that metastasizes to the infant. The original tumor in the placenta is rarely diagnosed because it is usually too small to be recognized on inspection [11]. Children with infantile choriocarcinoma become symptomatic at a median age of about 1 month [4]. Infants are often unstable due to hemorrhage. The diagnosis may be made without biopsy based on extremely high serum beta-hCG levels and normal AFP levels for age. It is a very rare tumor and only 32 cases have been described in the literature. Newborn infants tend to present clinically with a characteristic picture of anemia, hepatomegaly, hemorrhagic liver tumors, and precocious puberty Followed by rapid progression to death and maternal choriocarcinoma [12]. In 1968, Witzleben et al. described a constellation of clinical and laboratory findings that were seen in cases of "infantile choriocarcinoma syndrome": presentation at 5 weeks to 7 months of age, anemia and/or pallor, and hepatomegaly. A history of hemoptysis, hematemesis, melena, or hematuria was also often obtained. These infants were acutely ill [13]. Signs and symptoms of precocious puberty could also be present, related to the elevated levels of B-HCG. Although not specific for choriocarcinoma, these findings are suggestive and demand a follow-up [3,14].

Few studies have described the imaging appearance of infantile hepatic choriocarcinoma. Frequently the tumor is misdiagnosed as hemangioendothelioma, mesenchymal hamartoma, hepatoblastoma, metastatic neuroblastoma and undifferentiated sarcoma [9].

Imaging findings are non-specific. In most reports, the CT scan shows a large complex mass with indistinct cystic and hemorrhagic components with peripheral enhancement. The primary lesion or the metastatic lesions may show large vascular channels on CT as well as sonography with color and duplex Doppler [15]. The MR imaging appearance varies due to the presence or absence of hemorrhage as well as central necrosis with peripheral enhancement pattern with or without filling of the lesion on delayed images [12]. The periphery of the mass may show high signal intensity on T1-weighted images (T1WI) and T2-weighted images (T2WI) suggesting hemorrhagic components.

Choriocarcinoma usually has heterogeneous T1 and T2 signal intensity with large areas of central necrosis. T1 hyper-intense hemorrhagic areas are frequently encountered in addition to fluid filled cysts with high protein content that are T1 and T2 hyper-intense and these features are like ovarian choriocarcinoma [12,16]. When compared to uterine choriocarcinoma, the imaging features are similar except that uterine choriocarcinoma tend to be infiltrative rather than presenting with masses. Increased tracer uptake by the lesion or the metastatic deposits may also be seen on bone scans [9].

Prognosis of infantile cases is poor. As our patient, the cause of death is usually life threatening bleeding. Most of the infantile cases diagnosed during life have died either before treatment started or despite treatment. There are six case reports of infants who have survived [13].

Despite its highly malignant nature, this tumor is extremely chemo-sensitive. However, as there are no established protocols, the treatments have usually been based on germ cell tumor protocols [17]. Chemotherapy may be followed by surgical resection of the involved hepatic segments. The use of liver transplant in cases of unrespectable tumors has also been reported [11].

Conclusion

Although although infantile choriocarcinoma is an extremely rare hepatic tumor in children, whenever encountered with a case of a hepatic mass in an unstable neonate with a clinical picture of hemorrhage, choriocarcinoma should be considered in the differential diagnosis in addition to infantile hemangioendothelioma. The β -HCG is usually high. If this possibility is entertained, the prognosis for affected children can improve due to early diagnosis and initiation of appropriate treatment.

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