Hepatoblastoma in a neonate: a hypervascular presentation mimicking hemangiendothelioma

Introduction

The differential diagnosis of a primary hepatic mass in a neonate is usually limited to hemangiendothelioma, a benign tumor, and hepatoblastoma, a malignancy. Since hemangiendotheliomas are highly vascular tumors, the vascularity on imaging studies has been a primary diagnostic consideration [1]. Arteriovenous shunting occurring in this entity may result in a clinical presentation of congestive heart failure. Kasabach-Merritt syndrome, a consumptive coagulopathy, has also been associated with this benign tumor. Based on clinical and radiological assessment, medical treatment for hemangiendothelioma has sometimes been initiated without first obtaining a tissue diagnosis [2].

A neonate recently presented to our hospital with a hepatic mass that radiologically appeared to be a classic hemangiendothelioma. The presence of congestive heart failure and Kasabach-Merritt syndrome supported this diagnosis; however, surgical pathology proved the mass to be a hepatoblastoma.

Case report

A male neonate was transferred to our hospital on day 1 of life with a palpable abdominal mass and respiratory failure. Two weeks prior to delivery, the mother of the child noticed decreased fetal movement, which became even more pronounced 2 days prior to delivery. After an emergency cesarean section at 35 weeks’ gestational age, the baby was floppy, cyanotic, and apneic and required intubation. Apgar scores were 3 at 1 min, 5 at 5 min, and 9 at 10 min. Upon arrival at our hospital, physical examination revealed an intubated infant with diffuse rales on auscultation and a markedly distended abdomen with firm liver palpable to 8 cm below the right costal margin. No skin lesions were visualized. Urine output was decreased.

Laboratory evaluation revealed metabolic acidosis, thrombocytopenia (28,000/μl), PT 30 s (normal 16 s), PTT 43 s, fibrinogen < 15 mg/dl (normal 200–400 mg/dl), ALT 354 μm/l (normal < 54 μm/l), AST 1870 μm/l (normal 20–65 μm/l), and serum alphafetoprotein 50.372 ng/ml (normal < 215,166 ng/ml adjusted for a 35-week pre-term infant [3]). Echocardiogram showed reduced left ventricular function with a shortening fraction of 23%.

An abdominal ultrasound showed a large complex mass in the right lobe of the liver. The mass was inhomogeneous in echotexture with hypervascularity at the periphery of the mass and in the adjacent liver. The hepatic artery and hepatic veins were enlarged.

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Abstract

Congenital heart failure in the neonate supported by classic imaging findings may allow the implementation of medical therapy for presumed hemangiendothelioma without obtaining a tissue diagnosis. This case report describes a neonate with these classic clinical and radiographic findings but who underwent surgery for failing medical treatment and was diagnosed as having a hepatoblastoma by pathology. This case supports the need to obtain tissue confirmation before beginning medical therapy.

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Fig. 1 a Color Doppler shows markedly increased vascularity (arrow) in the heterogeneous mass. b The enlarged hepatic artery (HA) is similar in caliber to the aorta (AO).

Fig. 2 Cardiomegaly and pulmonary edema are signs of congested heart failure.

Fig. 3 a Noncontrast CT reveals central necrotic area in right hepatic lobe with calcifications (arrow). b Enlarged hepatic artery (arrow) feeds intensely enhancing mass replacing right hepatic lobe. c Hepatic veins (arrow) are also enlarged.