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Case Report

CASE REPORT

Rare cause of emergency in the first week of life: congenital hepatoblastoma (case report)

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Abstract

During the first week of life, a sudden deterioration in a newborn commonly includes investigations to rule out infections, lung pathologies, cardiac lesions, neurological insults, metabolic disorders or gastrointestinal emergencies. It is unusual, however, to consider malignancy as the primary causative factor. In this case report, we describe a rare and unusual presentation of congenital hepatoblastoma, its complications and management in a neonate with multi-organ dysfunction. A term infant presented with sudden deterioration, hemodynamic instability and an acute abdomen on his 4th day of life. Surgical exploration revealed a ruptured neoplasm that pathology diagnosed as a congenital hepatoblastoma. After the patient was stabilized, chemotherapy was initiated. At present, the patient is 8 months old and under continuous follow-up of oncology service. This case highlights the importance of considering rare diagnoses including congenital malignancy when investigating and managing a sick newborn with multi-organ dysfunction.

INTRODUCTION

Congenital hepatoblastoma (CH) is rare and often presents in a generalized and non-specific manner [1]. Therefore, it is much more challenging to consider and diagnose this condition in the neonatal population [1, 2]. In this case report, we describe a rare and unusual presentation of CH, its complications and management in a tertiary neonatal intensive care unit (NICU).

CASE REPORT

Our patient was a first child to the healthy, non-consanguineous parents. The pregnancy course was uneventful, and at 40 weeks of gestational age mother went into spontaneous labor. The baby boy was born via forceps-assisted vaginal delivery, with APGARS of 8 and 9 at 1 and 5 min of life, respectively. He was small for gestational age (weight of 2410 g), but his clinical assessment was otherwise unremarkable and was transferred to the postpartum unit for routine newborn care.

At around 15 h of life, he was noted to be hypothermic (core temperature of 36°C), but otherwise examined healthy. Accordingly, he was transferred to the NICU for further investigations where his complete blood count showed a normal white blood cells of ×15.1 10e9/L, mild polycythemia with hemoglobin of 185 g/L, platelets of 323 × 10e9/L, and a normal differential count. After obtaining a blood culture, he was started on empiric antibiotics. A TORCH screen was sent in view of growth restriction and was reported as negative. By 36 h of age, he was maintaining normal body temperature and tolerating full feeds. His septic work-up was negative, antibiotics were discontinued after 48 h, and the medical team was planning to discharge the baby home on the Day 4 of life.

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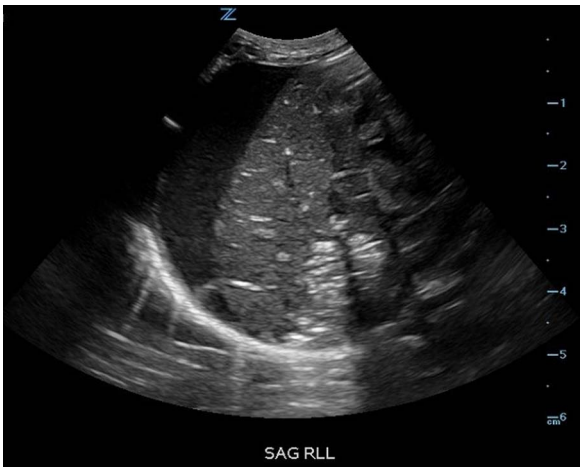

Figure 1: Fluids collection in an abdomen (hemoperitoneum).

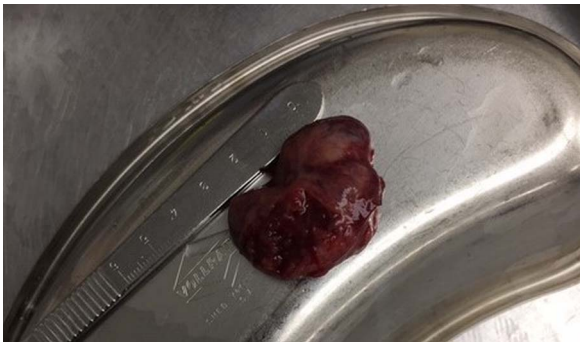

Figure 2: Resected liver mass.

On 4th day of life, his clinical status rapidly deteriorated with poor respiratory effort, hypotension and peripheral hypoperfusion, as well as abdominal distension and significant hepatomegaly. These changes initially were associated with short period of irritability and distress and were followed by lethargy and unresponsiveness requiring intubation. Urgent bedside cranial ultrasonography was unremarkable, as well as chest X-ray. Echocardiogram did not show any evidence of critical cardiac defects or obstructive lesions, but revealed very low intraventricular volume and thickened myocardium with decreased systolic function. He remained hypotensive in spite of extensive inotropic support and fluids management. An abdominal ultrasound showed a complex fluid collection in the abdomen, representing a possible hemoperitoneum (Fig. 1), and with increasing abdominal distension, he was taken to the operating room for an emergency laparotomy. A 3 × 8 cm mass arising from segment four of his liver was ruptured, bleeding and eventually resected after the intraabdominal blood was evacuated (Fig. 2). Malignancy was suspected and an oncologic investigation was initiated.

Post-operatively, he developed multi-organ dysfunction presenting as seizures, pulmonary hemorrhage, hypovolemic shock, acute kidney injury requiring continuous renal replacement therapy and disseminated intravascular coagulation. His alpha-fetoprotein was elevated (1940 u/L) and decreased significantly after the surgery (665 u/L) (Fig. 3). Magnetic resonance imaging (MRI) revealed the presence of at least four hepatic tumors (segments 4a, 4b, 7 and 8) that were T1 hypointense and T2 hyperintense (Fig. 4). The histopathology of the resected liver mass was reported as an epithelial type hepatoblastoma (mixed fetal and embryonal). After the diagnosis of hepatoblastoma, the goal was to stabilize patient for initiation of chemotherapy that was started at 6 weeks of age. Chemotherapy was tolerated well, and currently the patient is 8 months old and under continuous follow-up of oncology service.

DISCUSSION

During the 1st week of life, a sudden clinical deterioration is commonly caused by infections, lung pathologies, cardiac lesions, neurological insults, metabolic disorders or gastrointestinal emergencies. Malignancy, however, is very unusual but should be considered when investigating and managing an undifferentiated sick newborn.

Hepatoblastoma is rare in pediatrics, with <10% being diagnosed in the neonatal period [1]. The term 'congenital hepatoblastoma' is used when the liver tumor is detected antenatally and ~3 months of age. A 2007 retrospective review of congenital hepatic lesions identified hepatoblastomas as being the least common (16.5%) [2]. Out of all primary hepatic lesions, hepatoblastoma had also the worst outcome with a 25% survival rate and a 100% mortality rate for Stage 4 hepatoblastoma [2]. Of note, the 25% overall survival rate consisted of both fetal and neonatal patients as well as those who were both treated and untreated, depending on a combination of clinical stability, size of tumor and prognosis [2].

Antenatally, hydramnios and fetal hydrops can be indicators of CH [2]. During labor, mechanical compression of the tumor can cause the tumor to rupture and lead to intraabdominal hemorrhage. In the postnatal period, it is most often diagnosed clinically by the presence of a palpable abdominal mass commonly associated with abdominal distension and respiratory distress [2]. Associated features of hemihypertrophy, Beckwith-Wiedemann and intestinal polyposis [3] have also been noted. Low birth weight and prematurity [2] have been identified as risk factors. Of note, our patient was small for gestational age but had an unremarkable neonatal exam until 4 days of life where he developed hemodynamic instability and an acute abdomen.

Laboratory findings suggestive of CH include anemia [2], thrombocytopenia [2] or thrombocytosis [4] and an elevated alpha-fetoprotein in approximately half of the neonates [2]. Our patient had a normal platelet count and hemoglobin at birth, but during his clinical deterioration and intra-abdominal hemorrhage, he developed persistent thrombocytopenia and severe anemia. Although his alpha-fetoprotein was elevated, it is not a reliable indicator to diagnose CH [2] as it lacks specificity and is only elevated in approximately half of neonates.

Imaging studies often begin with abdominal ultrasounds showing a well-defined and heterogenous solid mass, with varying levels of cystic changes, calcifications and necrosis [3]. A 2011 review outlining the imaging methods and timing of congenital tumors identified the utility of imaging CH for evaluation the anatomical extent and relationship with hepatic lobar anatomy prior to surgical planning [3]. An antenatal diagnosis was confirmed in only 1/3rd of the cases, with most diagnoses being made in combination with a clinical exam and neonatal ultrasound after birth [2]. A 2011 case report and review highlighted the importance of antenatal diagnosis to improve patient outcomes [6]. Location of CHs were most often localized to the right lobe in >50% of the cases [2, 7].

Management of CHs generally involve complete surgical resection of the tumor, chemotherapy and possible liver

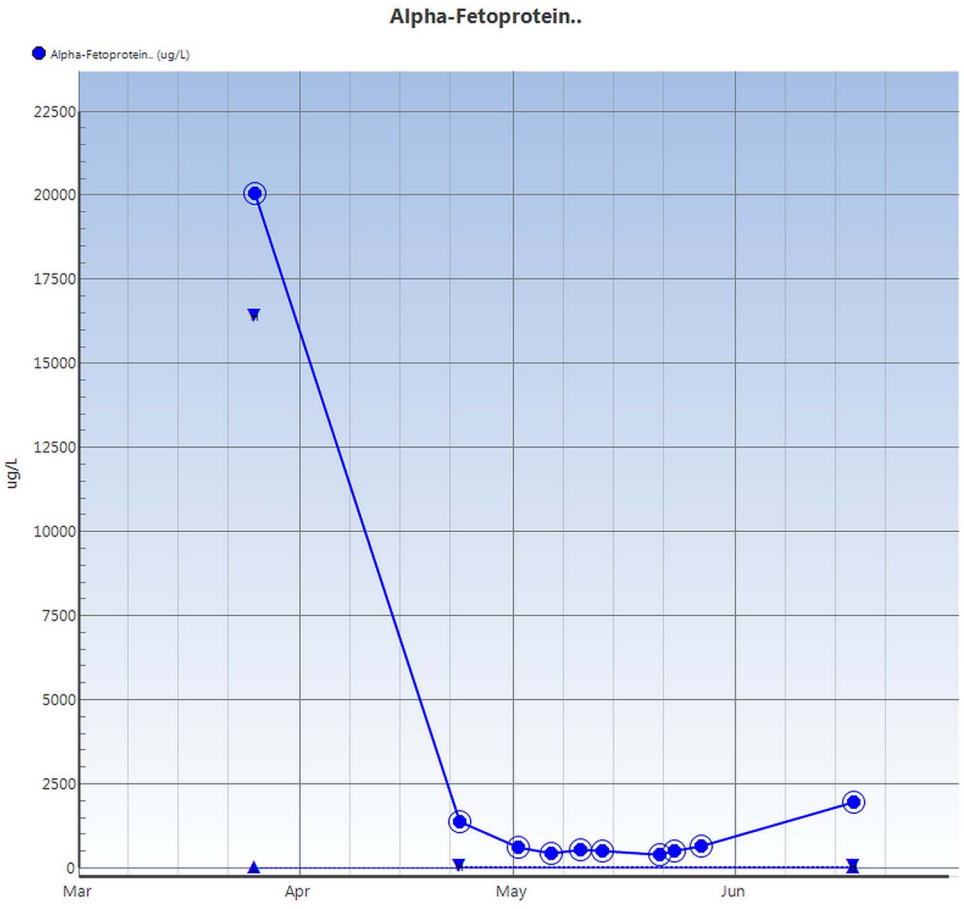

Figure 3: Alpha-fetoprotein level during clinical course.

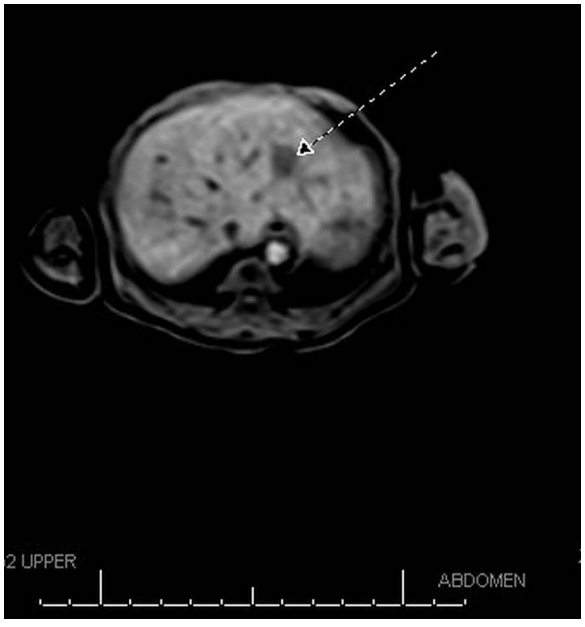

Figure 4: Post-surgery MRI showing intra-hepatic lesion (arrow).

transplant. A histological diagnosis is recommended to confirm the diagnosis and subtype of the tumor, with the majority being a mix of the fetal and embryonal type [2]. Left untreated, CHs are generally fatal by 2 years of age [2, 5, 7]. The main cause of postnatal death involves a mass effect leading to abdominal distension, compression of vasculature, severe respiratory distress and perioperative complications [2, 7]. Additionally, one study showed that all those patients in Stage 4 disease did not survive regardless of treatment modality [2].

CH is rare in the neonatal population and often not on the list of differential diagnoses for a sick neonate. This case highlights the importance of considering malignancies when investigating and managing a sick newborn with multi-organ dysfunction.

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CONFLICT OF INTEREST STATEMENT

None declared.

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ETHICAL APPROVAL

None required.

CONSENT

Informed written consent was taken from the patient' parents.

GUARANTOR

V.M.S. is the guarantor of this work.

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