Case Report

Congenital hepatoblastoma in a neonate

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ABSTRACT

Hepatoblastoma is a rare cause of mass abdomen and distension of abdomen in neonates. In this report, a 1-day-old baby who presented with abdominal distension at birth is being described for its unusual presentation. Diagnosis of hepatoblastoma in the antenatal period is possible with the available imaging modalities such as ultrasound abdomen. Caesarean section is recommended when HBL is antenatally diagnosed as vaginal delivery may cause tumor rupture. Beckwith-Wiedemann syndrome, hemihypertrophy and low birth weight are common associations of HBL. Serum Alpha feto protein (AFP) is the most useful laboratory test for hepatoblastoma and hepatocellular carcinoma. AFP is produced in the fetal liver and yolk sac, and levels decline to adult values during the first 6 months after birth. AFP can be used as a tumour marker for screening and confirmation of diagnosis though it is not a very specific tumour marker. Hepatoblastoma should be considered in the differential diagnosis when a neonate presents with distension of abdomen.

Keywords: Abdominal distension, Beckwith-Wiedemann syndrome, Hepatoblastoma

INTRODUCTION

Tumours detected antenatally or after birth up to 3 months of age are considered congenital fetal tumours. The lesions can be benign or malignant, and early detection of the tumor can significantly affect neonatal care. Hepatoblastoma is a rare primary malignant liver tumor affecting mainly pediatric patients in the age group 6 months to 3 Years and is rare in neonatal period. Congenital HBLs exhibit some important distinctive features when compared to HBLs diagnosed in children beyond the neonatal period.¹ These are: a different clinical presentation, a higher incidence of pure fetal histology, a significant risk for systemic metastases and a worse outcome In this report hepatoblastoma arising from the right lobe of liver presenting as massive distension of abdomen is being described in a day old neonate.

Antenatal detection using Ultrasound abdomen helps in proper planning of management of such cases.

CASE REPORT

A one day old, term AGA male baby was referred to our hospital presenting with abdominal distension since birth. An antenatal scan was done 15 days before birth, which showed an abdominal mass. On clinical examination, gross abdominal distension and prominent veins over abdomen were observed. Ultrasound and CT showed a big mass arising from right lobe of the liver likely hepatoblastoma. Laparotomy was done, mass was resected and sent for histopathological evaluation. The histopathological features were suggestive of hepatoblastoma wholly epithelial type with mixed fetal and embryonal sub types. Post surgery the baby could not be revived.
Hepatoblastoma (HB) is the most frequent malignant liver tumor in infants and young children, and it represents the third most common intra-abdominal pediatric malignancy following Neuroblastoma and Wilms’ tumor. They usually occur in children between the age of 6 months and 3 years.\(^1\)

Less than 10% of HBL cases are diagnosed in the neonatal period.\(^2\) Incidence of hepatoblastoma is 1.2 per million and in less than 1 yr is 1 per million.\(^3,4\) Tumors detected antenatally or after birth up to 3 months of age are considered congenital fetal tumors. The lesions can be benign or malignant, and early detection of the tumor can significantly affect neonatal care.\(^5\)

Congenital HBLs exhibit some important distinctive features when compared to HBLs diagnosed in children beyond the neonatal period.\(^6\) These are: a different clinical presentation, a higher incidence of pure fetal histology, a significant risk for systemic metastases and a worse outcome.\(^7\)

The etiology of HB has not been fully elucidated. Early studies have correlated HB occurrence with parental exposure to metals, petroleum, and paints and more recently, with adverse events associated to prematurity and low birth weight.\(^8,9\) It is sometimes found to be associated with congenital abnormalities and overgrowth syndromes including the Beckwith Weidman syndrome (BWS), hemihypertrophy, and occasionally trisomy 18, Prader-Willi, and Simpson-Golabi Behmel syndromes.\(^10\)

In HB, loss of maternal 11p15.5 alleles has been associated with overexpression of IGF2 and alteration of the IGF axis.\(^11,13\) The important role of IGF2 in liver development and its growth-promoting and anti-apoptotic activities strongly implicate this factor in HB pathogenesis.

In most cases, children present with abdominal distension, abdominal pain, and gastrointestinal disorders. Liver function tests such as alanine aminotransferase, total bilirubin, albumin, and alkaline phosphatase remain generally normal. Elevated level of serum AFP represents a fairly constant marker, as it has been found in 90% of HB cases. However, AFP levels must be interpreted with caution because AFP is commonly elevated in normal neonates until 6 months of age, and it may be elevated also in association with other tumors including hemangiomas/ hemangioendotheliomas and mesenchymal hamartoma of the liver.

Right lobe of liver is more commonly found to be affected by HBL, as was seen in our case. This could be related to the different blood supply of the two lobes, with low oxygen tension of portal supply to the right lobe in contrast to umbilical venous supply to the left lobe having some role in the embryological differentiation of HBL.\(^14\)

Early detection of HBLs allows for consideration of the mode of delivery, prompt treatment, and may lead to better outcomes.\(^6\) Caesarean section is recommended when HBL is antenatally diagnosed as vaginal delivery may cause tumor rupture.\(^15\)

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REFERENCES
