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CASE REPORT

HEPATOBLASTOMA IN A LOW BIRTH WEIGHT INFANT : A CASE REPORT AND REVIEW OF THE LITERATURE

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Hepatoblastoma is the most common primary liver tumour of childhood. This is a case report of a one-year-old boy who presented with a one-month history of progressive abdominal distension and weight loss. He was cachexic, anaemic, had gross hepatomegaly and ascites. He had been born prematurely with a birth weight of 1.23 kg, and his developmental milestones were delayed. Ultrasound and CT scan demonstrated a large solid tumour in the left lobe of the liver with a smaller superficial nodule in the right lobe. Serum alpha fetoprotein was significantly raised. A left lobe hepatectomy and complete excision of the right sided nodule was performed. There was no evidence of metastatic disease. Histopathological examination confirmed hepatoblastoma of the fetal type. The patient developed features of intestinal obstruction a few days after surgery and he succumbed ten days after re-laparotomy. The clinical presentation and investigation results in this case are characteristic. Recent reports have suggested a strong relationship between very low birth weight (< 1500gm)/prematurity and hepatoblastoma as is present in this case. Surgery is the mainstay of therapy in hepatoblastoma. A brief review of the literature on this tumour is presented.

Key words : *hepatoblastoma, low birth weight*

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Introduction

Hepatoblastoma accounts for 25-40% of paediatric liver tumours. The incidence throughout the world is fairly constant at 0.5 – 1.5 cases per million children (1). Statistics from the medical record department in our institution revealed no cases of primary liver tumours in children in the last ten years.

Case Report

A one-year-old Malay boy presented with a one-month history of progressive abdominal distension and weight loss. He was cachexic, anaemic, had gross hepatomegaly and ascites. There were no stigmata of chronic liver disease.

He had been born prematurely at 28 weeks of gestation with a birth weight of 1.23 kg. His developmental milestones were delayed.

Laboratory investigations revealed a haemoglobin of 9.9g/dL and thrombocytosis. His serum alpha fetoprotein (αFP) level was above 1000mg/mol. The liver function tests were normal and there was no evidence of hepatitis B and C infections.

Ultrasound and CT scan demonstrated a large solid tumour in the left lobe of the liver measuring 12 x 11 x 8 cm with a smaller superficial nodule in the right lobe. There was no evidence of metastatic disease outside of the liver.

A left lobe hepatectomy and complete excision of the right sided nodule was performed. The draining lymph nodes appeared normal.

Gross pathological examination of the left lobe tumour (Figure 1) showed an oval soft mass of

500g and 12 x 10 x 7 cm in dimensions. It had a pseudocapsule and cut section was gray and bile-stained with foci of necrosis. The right sided nodule had similar features.

Microscopic examination revealed a well-circumscribed, non-encapsulated tumour composed of uniform immature epithelial cells resembling fetal hepatocytes. They were organised in cords and sheets with a sinusoidal pattern. Foci of extramedullary haemopoiesis was noted. Immature mesenchymal elements were absent (Figure 2). The tumour cells stained positively for αFP.

The patient developed features of intestinal obstruction a few days after surgery which was attributed to paralytic ileus after a re-laparotomy.

Unfortunately the child's condition worsened and he succumbed ten days after laparotomy due to septicaemia.

Discussion

The median age of onset of hepatoblastoma is approximately 16 months with a slight male predominance.

The cause is unknown. It is considered to be an embryonal tumour that is related to developmental disturbances of cell proliferation and differentiation during hepatic organogenesis (1,2).

Most cases are sporadic. Hepatoblastoma has been reported in association with other disorders including Beckwith-Wiedeman syndrome, familial adenomatous polyposis, Wilms tumour and other congenital anomalies such as hemihypertrophy and cleft palate (2). These were not evident in this case.

Some risk factors that have been implicated in hepatoblastoma include oral contraceptive intake, alcohol abuse during pregnancy, hormonal treatment, maternal occupational exposure to metals, petroleum products and paints (2). Several recent papers have noted an association between hepatoblastoma and very low birth weight infants (< 1500g)/ prematurity as is present in this case (1-4). There is a significant correlation between the gestational age and the stage of disease whereby hepatoblastomas with unfavourable biologic behaviour and advanced stage developed in children who were extremely premature at birth. In this case the gestational age of 28 weeks and birth weight of 1.23 kg corresponded with the stage II tumour (tumour involving two segments or infiltrating beyond tumour capsule) as reported by Ikeda H et al (2). It is hypothesized that genetically altered liver cells exist in some premature infants and the various procedures and treatments received by these infants cause tumour promotion or transformation (3-4).

The clinical presentation of a growing abdominal mass associated with weight loss are characteristic. Other presenting features are anorexia, vomiting and failure to thrive. Anaemia and thrombocytosis are commonly seen at diagnosis.

Figure 1. Macroscopic picture of the left hepatic lobe tumour .

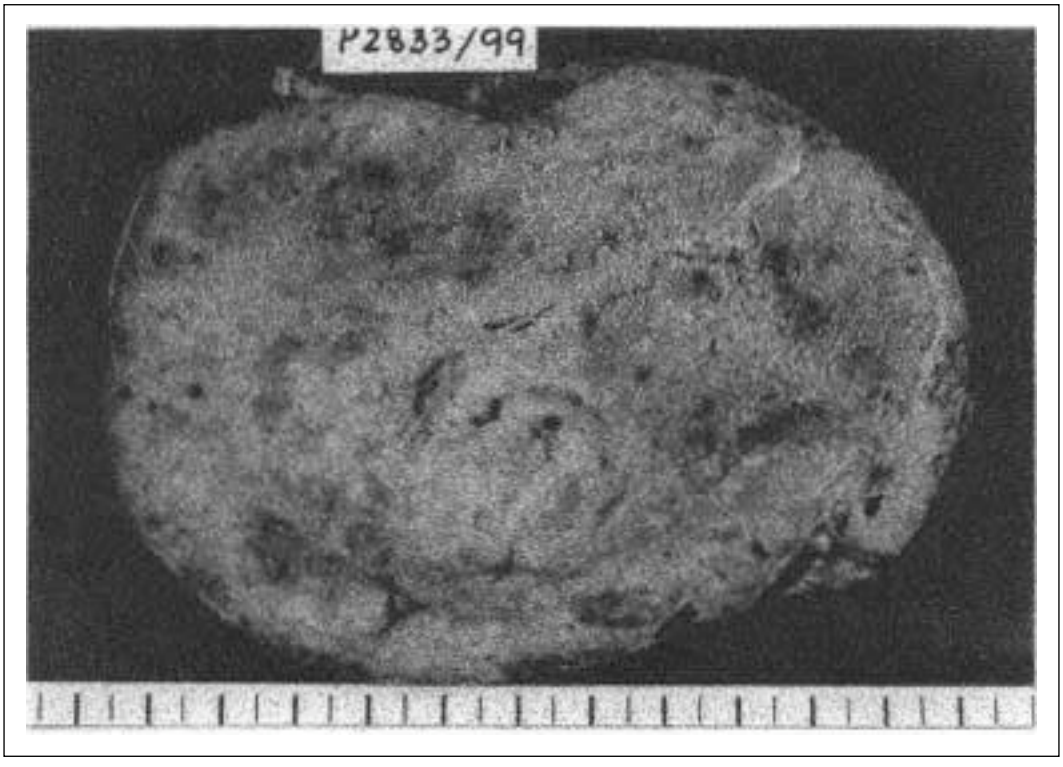
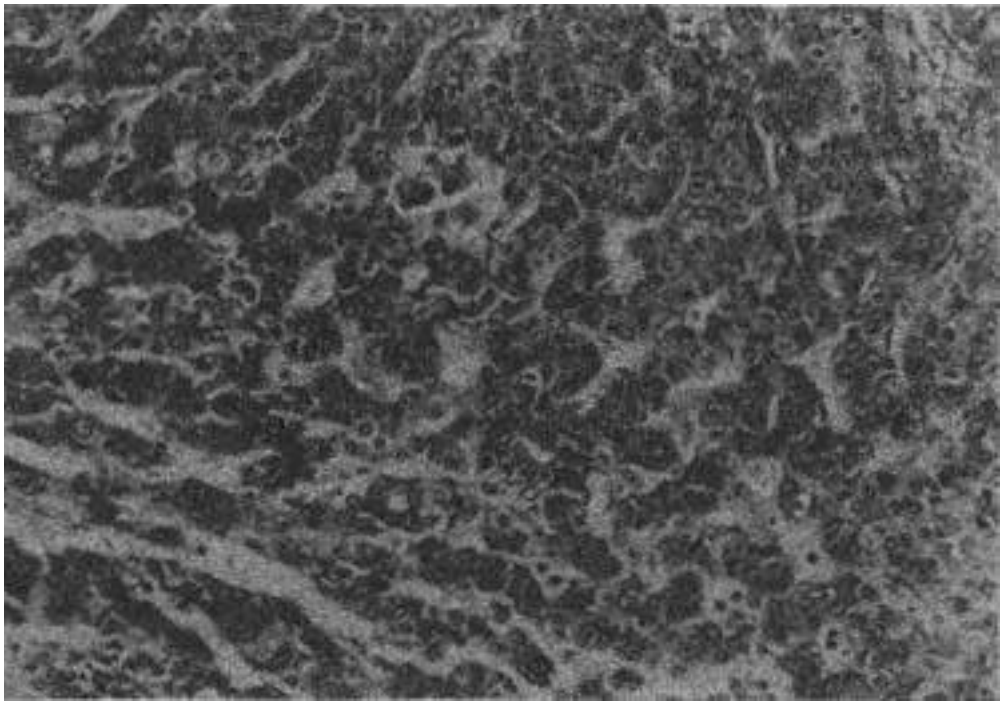


Figure 2. Microscopic picture of hepatoblastoma (fetal type), H&E, X 100.



An elevated serum αFP is found in more than 75-90% of cases of hepatoblastoma. Human chorionic gonadotrophin may also be produced by some tumours.

Hepatoblastomas are usually solitary (80%) and situated in the right lobe of the liver though this patient had a multifocal tumour. Its diameter ranges from 5 to 25 cm. Its cut surface of tan to bile-stained with necrosis are usual. Histologically hepatoblastoma is most often composed of epithelial hepatocytic elements with different subtypes, of which the fetal type is the most common. The histological pattern demonstrated in this case is typical of the fetal type. The other subtypes include embryonal, macrotrabecular, anaplastic and mixed (epithelial and stromal components). Immunohistochemically, reactivity for αFP with variable expression of other markers such as EMA(epithelial membrane antigen), cytokeratin and vimentin is observed.

A combination of the clinical findings of young age (below 3 years old), elevated serum αFP, thrombocytosis and an intrahepatic abdominal mass is considered almost pathognomonic of hepatoblastoma (1).

Hepatoblastoma is an aggressive neoplasm that invades locally and eventually metastasizes to regional lymph nodes, lungs, brain and other organs.

The most important prognostic factor is tumour stage at presentation and resectability. Histologic subtypes are less important although macrotrabecular and anaplastic variants are considered to have a worse outcome.

The treatment of choice is surgical resection of the tumour. Chemotherapy reduces tumour volume and the response rate to cisplatin-containing regimes varies from 70 – 90%. Hence some study groups recommend preoperative chemotherapy, deferring definitive surgery until after 2 to 3 months of therapy. Overall, the 3 year survival rate is 62-70% regardless of the first therapeutic modality used (1).

It has been suggested that children weighing < 1500g at birth, especially those born at 23 – 25 weeks of gestation should be monitored using serum αFP measurement and abdominal ultrasonography after discharge from the neonatal intensive care unit as they are at high risk of developing advanced hepatoblastoma (2).

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