



Case Report

Hepatoblastoma in adult age. A case report and literature review

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Abstract

Hepatoblastoma (HB) rarely occurs in adults. We report herein the unusual case of a 19-year-old, otherwise healthy woman with no history of liver disease who presented with upper abdominal pain and hepatomegaly. Tests for hepatitis B virus (HBV), hepatitis C virus (HCV) were negative, and AFP was normal. There was no evidence of liver cirrhosis. A well-demarcated solid mass of 14 cm in diameter, which was lobulated and partly necrotic, was detected in the liver by computed tomography (CT). At surgical exploration a large liver mass was detected occupying the entire right lobe. A right trisegmentectomy was performed with tumor grossly resected with microscopic residual disease (i.e positive margins). On microscopic examination the tumor was composed mainly of two components which were intermingled: epithelial and mesenchymal elements. The epithelial component was formed of small embryonal cells, grouped into nodules, scattered in cellular mesenchymal tissue. The diagnosis was mixed hepatoblastoma. The patient received 4 cycles of systemic chemotherapy with cisplatin and adriamycin. Post-chemotherapy evaluation revealed recurrence of the hepatoblastoma in the remaining liver. She died 6 months later.

Key words: Hepatoblastoma, adult age, liver neoplasm, chemotherapy.

Introduction

Hepatoblastoma is a rare malignant tumor of the liver and usually occurs in the first three years of life.¹ Most of these tumors arise in the embryo; hence it seems to be unusual that hepatoblastomas occur in adults and are an exceptional cause of primary malignant liver tumor in adult age patients.² Various synonymous has been used to describe this kind of tumor such as hepatic embryonic mixed tumor, rhabdomyosarcohepatoma, carcino-osteochondromyxosarcoma and malignant mixed hepatoblastoma.⁴⁻⁶ Since Bartok⁷ in 1958 described the first hepatoblastoma case in an adult patient, less than 40 cases have been reported in the literature.^{2,8-12} We report herein the unusual case of a 19-year-old, otherwise healthy woman with no history of liver disease who had a mixed hepatoblastoma.

Case report

A 19 year-old white female was referred to our hospital, with a 4 month history of epigastric pain and a palpable mass in epigastric area. She denied use of drugs or contraceptives. Physical examination revealed a palpable liver, 8 cm below the right costal margin. Laboratory studies showed: AST 53 U/L, ALT 41 U/L, alkaline phosphatase 238 mg/L, alpha-fetoprotein 2.5 ng/mL, calcium 12.7 mg/dL, phosphorus 3.6 mg/dL. Tests for hepatitis B virus (HBV) and hepatitis C virus (HCV) were negative. There was no evidence of liver cirrhosis on either the laboratory or imaging examinations. Abdominal CT scan revealed a 14 x 12 x 10 cm low-density mass in the right lobe of liver.

Magnetic resonance imaging (MRI) showed a large right hepatic lobe mass (*Figure 1*). Selective celiac arteriogram revealed that the mass was hypervascular (*Figure 2*). At surgical exploration a large liver mass was detected occupying the entire right lobe. A right trisegmentectomy was performed with tumor grossly resected with microscopic residual disease (i.e positive margins). On microscopic examination the tumor was composed by a combination of mesenchymal and epithelial elements. The diagnosis was mixed hepatoblastoma (*Figure 3*).

The postoperative course was uneventful and calcium level dropped to 7.8 mg/dL. The patient received 4 cycles of systemic chemotherapy with cisplatin and adriamycin.

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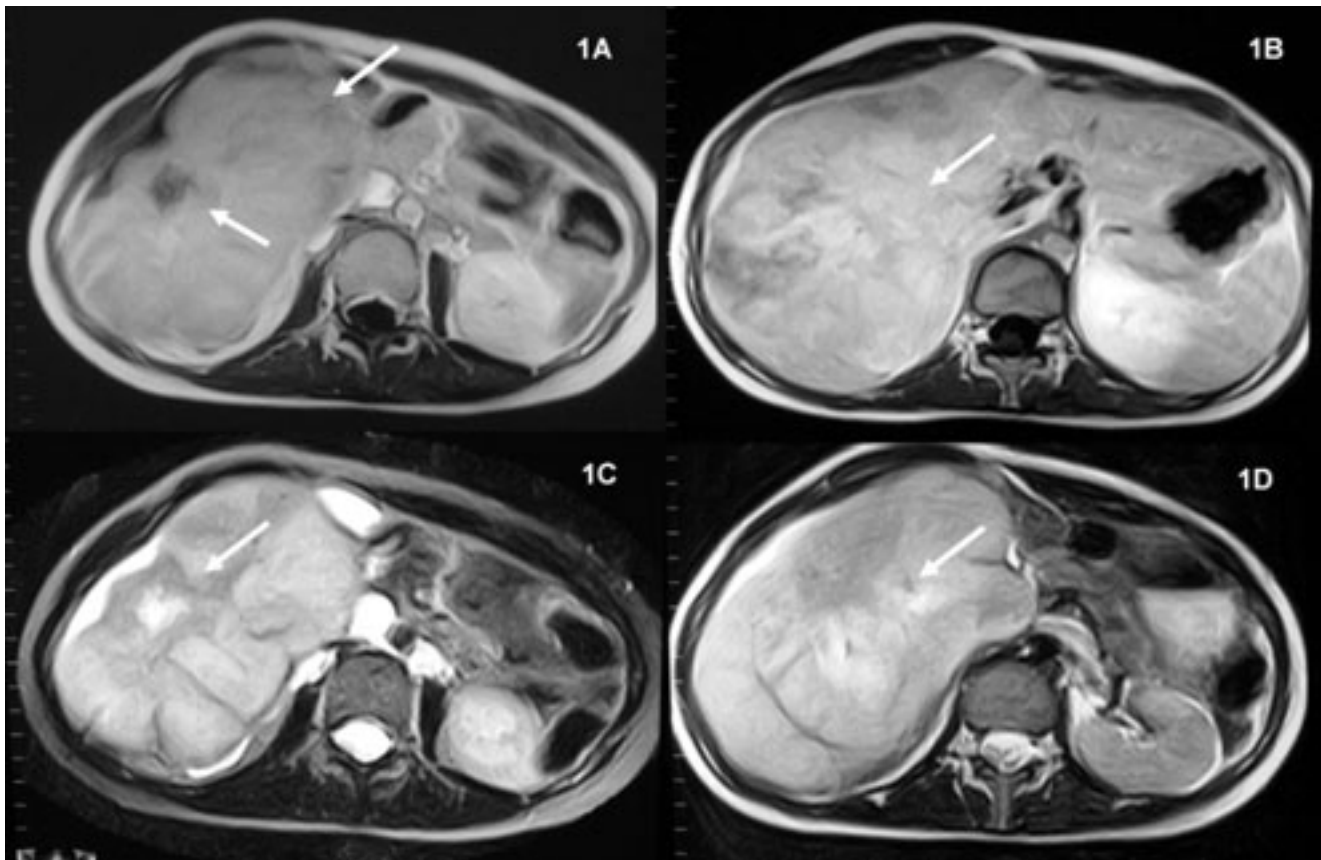


Figure 1. Magnetic resonance imaging (MRI) images. MRI showed on unenhanced T1-weighted transverse image a right hepatic lobe mass (arrows) that is hypointense. **A.** Enhanced T1-weighted transverse image of the liver obtained 3 minutes after administration of gadolinium shows a slight, unhomogeneous contrast material uptake. **B.** Unenhanced T1-weighted fat-suppressed transverse image reveals areas of hyperintensity indicating hemorrhage. **C.** On the T2-weighted transverse image the lesion is slightly hyperintense in comparison to remaining liver parenchyma **D.**



Figure 2. Selective celiac arteriogram. A hypervascular mass is noted.

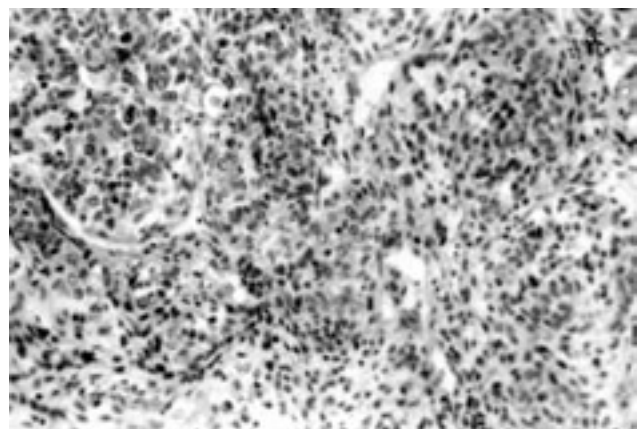


Figure 3. Histological findings. The tumor was composed mainly of two components which were intermingled: epithelial and mesenchymal elements. The epithelial component was formed of small embryonal cells, grouped into nodules, scattered in cellular mesenchymal tissue. (Haematoxylin and eosin).

cin. Post-chemotherapy evaluation revealed recurrence of the hepatoblastoma in the remaining liver. She was considered for orthotopic liver transplantation in our institution, but she died 6 months later.

Discussion

Hepatoblastomas accounts for 0.2-5.8% of total malignancies of the liver and for 25%-45% of primary hepatic

tumors in childhood.¹³ Approximately 90% of the cases occur in patients under 5 years of age and two thirds of the cases occurs in the first 2 years of life.¹³ Hepatoblastomas in adolescent and young adults is extremely rare and the prognosis is poor because they are usually diagnosed late. Initial symptoms are non-specific and the usual presentation is failure to thrive, loss of weight and a rapidly enlarging abdominal mass. In our case symptoms, physical and radiological findings were not different from those of the usual hepatocellular carcinoma.

Histologically, hepatoblastomas may present in two variants: a) the epithelial type, which consists of fetal and embryonic cells presenting alone or in combination; b) the epithelio-mesenchymal mixed type, in which mesenchymal elements are present along with the epithelial component.^{14,15} For a long time it has been thought that hepatoblastoma develops during intrauterine life, but the same histological pattern has been seen in hepatic tumors in adults, and new data about its histogenesis are emerging. An interesting theory looks at the common hepatocytes as the starting point, after having lost the differentiation and acquired new possibilities of transformation.⁹ As reported previously in childhood, absence of elevated alpha-fetoprotein, as in our patient, may be a poor prognostic factor.¹⁶ Occasionally hepatoblastomas produce beta-human chorionic gonadotropin resulting in isosexual precocity.¹⁷ Severe osteopenia is not uncommon. Hepatoblastoma is part of the constellation of findings associated with the Beckwith-Wiedemann syndrome.¹⁷ Hypercalcemia as in this case, was not reported previously.

Surgical resection is the cornerstone of treatment for patients with hepatoblastoma. Between 1990 and 1994, the Society of Pediatric Oncology Liver Tumor Study Group launched its first prospective trial (SIOPEL-1) with the intention to treat all patients with preoperative chemotherapy and delayed surgical resection.¹⁸ This study revealed that preoperative chemotherapy (cisplatin and doxorubicin) seems to make tumor resection easier. Resection of a positive resection margin does not necessarily have to be performed, because postoperative chemotherapy showed good results. According to the post surgical staging our patient was in a stage II. In children with stage I and II, surgical resection and systemic chemotherapy have a cure rate of 60%-90% approximately.¹⁹

Because the lack of experience of hepatoblastomas in adult patients, we think that those tumors can be treated with surgical resection (if it is possible) and chemotherapy as in children. However, mean survival time in adult patients is 3.5 months. Liver transplantation has recently been associated with significant success in the treatment

of children with unresectable hepatic tumors.¹⁹ Post transplant survival rates as high as 80% have been reported for children with hepatoblastomas.¹⁹ Intravenous invasion, positive lymph nodes and contiguous spread did not have a significant adverse effect on outcome. For children with hepatocellular carcinoma the overall five-year disease-free survival rate was approximately 60%. In conclusion, hepatoblastoma in adult patients has an aggressive presentation and a poor prognosis compared than childhood patients.

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