Hepatoblastoma in an Adult with Chronic Hepatitis

SHAW-NAN JEAN¹ XIAN-JIA JIANG¹ YUNGFANG CHEN² JUI-FEN CHEN² CHUNG-CHENG YEN¹

Department of Radiology¹, DA Chien General Hospital, Miaoli, Taiwan
Department of Radiology², China Medical College Hospital, Taichung, Taiwan

ABSTRACT

Hepatoblastoma usually occurs in children, but a few cases have also been reported in adults. We report an unusual case of hepatoblastoma in a 38-year old male with chronic hepatitis B infection. He visited our emergency room with intermittent right upper abdominal pain. At admission, elevated liver function tests were found. A test for hepatitis virus revealed that he has hepatitis B infection with marked elevation of alpha-fetoprotein (AFP) and up to 150593 IU/ml. abdominal ultrasound showed a 6.4x4.2 cm mass with mosaic pattern in the left lobe of the liver. Abdominal computed tomography (CT) demonstrated a large low-density mass occupying the left lobe and portal vein thrombosis. Pathological examination showed a hepatoblastoma. The patient received systemic chemotherapy with cisplatinum and adriamycin. Post-chemotherapy evaluation revealed poor response to this treatment. He died a few months later.

Correspondence Author to: Shaw-Nan Jean
Department of Radiology, DA Chien General Hospital, Miaoli, Taiwan
No. 6, Shin-Guang Street, Miaoli 360, Taiwan
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Abdominal computer tomography demonstrated a large, low-density mass without enhancement occupying the left lobe of the liver (Fig. 2). Tumor showed no early enhancement in arterial phase. Portal vein thrombosis from bifurcation of the left branch was found. Biopsy with histological examination showed a highly differentiated hepatoblastoma. The picture shows needle shaped fragments of inflammatory fibrous tissue with infiltrating individuals and vague clusters and plates of poor differentiated (embryonal-like) neoplastic cells (Fig. 3). The immunobiochemical study showed that Glypican-3 (GPC 3) is immunopositive in cytoplasm of the tumor cells (Fig. 4). Glypican-3 is a heparin sulfate proteoglycan bound to the cell surface that results from embryonic cell growth and differentiation. This case was not suitable for surgical intervention because of the portal vein invasion, so the patient received systemic chemotherapy with cisplatinum and adriamycin. Post-chemotherapy revealed poor response to treatment. He died a few months later.

DISCUSSION

Hepatoblastoma accounts for 0.2-5.8% of total malignancies of the liver and for 25%-45% of primary hepatic tumors in childhood [5]. Approximately 90% of cases occur in patients under 5 years of age and two thirds of the cases occur in the first 2 years of life [5]. Hepatoblastoma in adolescent and young adults are extremely rare, and the prognosis is poor because they are usually diagnosed in late stage. 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, the symptoms, physical and radiological findings were not different from those of the usual hepatocellular carcinoma (HCC). Most HCC are hypoattenuating on precontrast scans, and is frequently seen as a transiently hyperattenuating mass during the hepatic arterial phase. In the portal venous phase of enhancement, HCC may become isoattenuating with hepatic parenchyma. Although HCC is associated with chronic liver inflammation in adults, this association is not clear in adult hepatoblastoma. Our patient was a hepatitis B carrier. Because of the rare incidence of this tumor in adult, the initial consideration was a primary HCC associated with chronic hepatitis. However, radiological and blood chemical examinations revealed atypical findings of HCC, especially an extremely high AFP level. It has been shown that chronic hepatitis or liver cirrhosis is often present in adult cases, whereas in children hepatoblastoma can occur in a normal liver [6]. Although HCC arise in association with chronic liver disease, a similar etiological relationship might be possible in adult hepatoblastoma.

Histologically, hepatoblastoma 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 [7]. Mixed hepatoblastoma is common in adult cases.

Ultrasound is a noninvasive modality, particularly useful in the evaluation of infants. Hepatoblastoma is seen as a hyperechoic, solid, intrahepatic mass on ultrasound [8]. Ultrasound showed a hyperechoic mass with mosaic pattern in the left lobe of the liver in our case. Other standard investigations include computed tomography (CT), magnetic resonance imaging, and serum alpha-fetoprotein (AFP). The CT appearances of hepatoblastoma generally have a density lower than that of normal hepatic parenchyma on unenhanced and contrast-enhanced scan. This tumor is often heterogeneous because it contains hemorrhage, necrosis or focal steatosis. Calcifications occur in approximately 50% of hepatoblastoma. Portal vein invasion is also common. In our case, CT scan demonstrated a large, low-density mass occupying the left lobe of the liver. Thrombosis of the umbilical portion of the left portal vein was also noted. Hepatoblastoma is hypointense with respect to
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The liver on T1-weighted MR images and hyperintense on T2-weighted sequences. On post-contrast images, hepatoblastoma demonstrates diffuse, heterogeneous enhancement. However, the final diagnosis relies on tumor biopsy.

AFP is an extremely important tumor marker for the diagnosis of liver tumors in infants and evaluation of the effectiveness of treatment. AFP is positive in 90% of hepatoblastoma in infants and its positivity is higher than in adult HCC [9]. In adults, AFP level is relatively low as compared with infants [6]. In the present case, serum AFP was markedly elevated. In cases where AFP is positive, it would be a good indicator for the patient’s follow-up and detection of tumor recurrence.

The treatment for hepatoblastoma involves surgical resection and chemotherapy. Complete surgical resection is the cornerstone of treatment for patient with hepatoblastoma and is the only chance of an optimal clinical result. Despite this, the improvements in survival that have occurred over the last three decades have been the function of standardized chemotherapy that reduces tumor size and enables complete tumor excision [10]. However, the prognosis in adults is very poor, with survival usually less than 1 year because most adult cases are unresectable [11, 12]. Combination chemotherapy with adriamycin and cisplatinum has

Figure 2. Abdominal computer tomography (CT): axial arterial phase a, axial venous phase b, Coronal venous phase c. Demonstrated a large, low-density mass occupying the left lobe of the liver (arrow). Thrombosis of the umbilical portion of the left portal vein was noted as well (arrowhead).
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been described as effective treatment for this kind of tumor [12, 13]. Liver transplantation has recently been associated with significant success in the treatment of children with unresectable hepatic tumor. Post-transplant survival rates as high as 80% have been reported for children with hepatoblastoma. Intravenous invasion, positive lymph nodes and contiguous spread did not have a significant adverse effect on outcome [7]. For children with hepatoblastoma, the overall five-year disease-free survival rate is approximately 60%. In conclusion, hepatoblastoma in adult patients has an aggressive presentation and carries a poor prognosis compared with that of children.

REFERENCES