Adult hepatoblastoma: Case report with adrenal recurrence

Dear Editor,

Hepatoblastoma is the most frequent primary liver tumor of childhood, with an annual incidence of 0.5-1.5 cases per million (1). It is very rare in adults, with a controversial existent since some authors believe that most of them are really underdiagnosed hepatocarcinomas, mixed sarcomas or hepatic cholangio-hepatocarcinomas. To date, there are only 40 reported cases of hepatoblastoma in adult patients. The first case was described by Barnett in 1958. (2) The only available treatment that has improved survival is radical surgery (R0). The main difference with children is the frequency of lymphatic or hematogenous metastases, which are extremely rare in the pediatric population (3,4).

Case report

We report the case of a 27-year-old male, born in Guinea, with chronic B hepatitis (HBsAg +, HBeAb +, HBeAg-, and HBeAg-), who was diagnosed with a large hepatic tumor of 17.2 x 15.3 x 22.2 cm replacing the entire right hepatic lobe. CT-scan showed a neoplasm with arterial contrast enhancement and, in the laboratory test, we found an alpha-fetoprotein of 1,166,000 ng/ml. Right hepatectomy extended to segment IV for a free resection margin (R0) was performed. It was a solid and yellowish tumor, with multinodular appearance and myxoid and necrotic areas. Histopathological study revealed the presence of a malignant primary liver tumor with features suggesting a mixed hepatoblastoma with fetal and embryonic areas. The immunohistochemical study confirmed the diagnosis. The patient received adjuvant chemotherapy according to SIOPEL 4. After 15 months follow-up, a PET-CT detected the presence of left adrenal recurrence. Total adrenalectomy was performed with complete resection of the tumor (R0). After 12 months follow-up of the second surgery, the patient has no further signs of recurrence.

Discussion

Adult hepatoblastoma has higher risk of metastases, synchronous or metachronous, that children hepatoblastoma (5). This is

![Fig. 1. A. Large hepatic tumor replacing the hepatic right lobe. B. Transection line during the surgery. C. Surgical hepatic specimen. D. Left adrenal recurrence. E. Anatomical relations of the adrenal recurrence. F. Surgical adrenal specimen.](image-url)
the second case reported in the literature that describes an adrenal recurrence after hepatoblastoma resection (6). The experience in the treatment of hepatoblastoma metastases is poor, and there are not well-defined protocols. To date, radical surgery (R0) of the metastases seems to be the only available curative treatment (7,8).

Hepatoblastoma is a typical tumor from children. Although some authors question the existence in adults, several cases have been reported in the literature. The prognosis is generally poor and the only available treatment is radical surgery (R0), both the primary tumor and metastases.


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References