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Case Report

Hepatoblastoma in an 18- Year Old Female; Rare Entity

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ABSTRACT

Hepatoblastoma is a rare malignant hepatic tumor in adults. It is associated with poor prognosis as it is usually diagnosed late when the tumor is completely unresectable. Presented here, is a case of an 18-year old female with pain abdomen, vomiting, and mass over the right hypochondriac region for one month. CT scan revealed soft tissue density mass on the right lobe of liver suggestive of focal nodular hyperplasia with differential diagnosis of atypical hemangioma. Liver angiography also suggested a vascular tumor of giant hemangioma with inferior exophytic extension. The patient underwent a right extended hepatectomy. Histopathological examination revealed hepatoblastoma with predominant fetal pattern and small focal areas of embryonal pattern. The patient had an uneventful postoperative recovery and is currently undergoing chemotherapy. We present this case for its rarity and ability to masquerade other primary liver tumors in the adult age as seen in our patient.

Keywords: Embryonal; Fetal; Hepatoblastoma, Hepatectomy

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INTRODUCTION

Hepatoblastoma is a tumor of the pediatric age group and accounts for 1-4% of all primary malignancies in children.[1] It is the most frequent malignant liver tumor in infants under three years of age.[2] It is postulated to arise from primary hepatoblasts or multipotent hepatic progenitor cells with the capacity to differentiate along with a variety of lineage. Since it arises in the embryo, hepatoblastoma is very rare in adults with only sixty cases reported in the literature so far.[3] In adults, it is related to a poor prognosis with a median survival of < 5 months.[4] Although surgery offers the only chance for a cure, it often recurs even after complete resection. Rougemont et al described a total of 45 cases of adult hepatoblastoma, all with dismal prognosis due to late presentation and with no consensus on the treatment due to the rarity of the disease.[5] We present a case of hepatoblastoma in an 18-year-old female.

CASE REPORT

It was a case of an 18-year-old female who presented with nonspecific symptoms such as pain abdomen over the right hypochondriac region and vomiting for one month. Weight loss and jaundice were not present. On abdominal palpation, a diffuse non-tender lump was palpable over the right hypochondriac region. Her CBC was within normal limits. The liver function test was deranged with elevated SGPT, SGOT, ALP, and Gamma GT. Serum AFP was raised (>520 ng/ml). Another tumor marker serum CA 19.[9] was also raised (>71.9 U/ml). However, CEA was within normal limits. USG showed hepatomegaly with hyperechoic lesion measuring 10.8x8.2 cm with few hypoechoic areas with increased vascularity. CT scan showed soft tissue density mass on the right lobe of the liver likely to be focal nodular hyperplasia. Differential diagnosis was atypical hemangioma. Liver angiography showed well defined heterogeneously enhancing mass measuring 15x14x10 cm with features suggestive of giant hemangioma with inferior exophytic extension. She underwent a right extended hepatectomy with cholecystectomy. She had received transarterial embolization of the right hepatic artery involving anterior and posterior branch two days prior to the surgery.

Right extended hepatectomy and cholecystectomy specimen was received in the Department of Pathology. The specimen consisted

of a liver measuring 16 x 16 x 8.5 cm with gall bladder measuring 7 x 3 x 0.2 cm. Cut section showed a large solitary circumscribed mass measuring 12 x 9 x 6 cm involving almost the entire liver specimen. The mass was solid grey white, soft and friable with extensive areas of necrosis and hemorrhage. On microscopic examination, the tumor was predominantly composed of fetal cells arranged in a nest and thin trabeculae showing characteristic alternating light and dark pattern. These cells were oval to polygonal in shape with abundant clear to eosinophilic cytoplasm, centrally placed oval nucleus, vesicular chromatin, and prominent nucleoli. Mitotic figures were infrequent (fig 1). In addition, multiple small focal areas of the embryonal pattern were noted. These areas showed nests and lobules of tumor cells with scant cytoplasm, high nuclear-cytoplasmic ratio, and moderate to marked nuclear pleomorphism (fig 2). Extensive areas of tumor necrosis were present (fig.3). Mesenchymal components were not seen. Focal capsular and vascular invasions were noted. Adjacent hepatic parenchyma was infiltrated by the tumor. The final diagnosis of epithelial hepatoblastoma, mixed fetal, and embryonal subtype was reported.

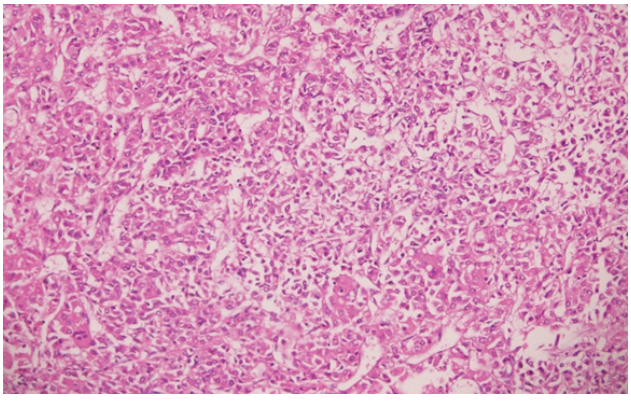

Figure 1: Photomicrograph showing fetal fetal cells arranged in a nest and thin trabeculae showing characteristic alternating light and dark pattern. (HE stain, X 100)

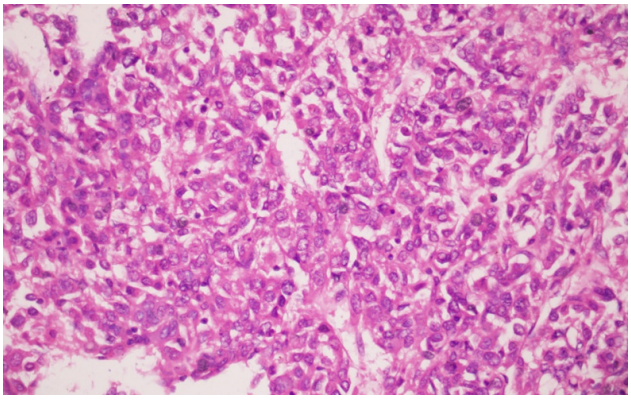

Figure 2: High power view showing embrynal pattern of cells with scant cytoplasm, high NC ratio, and nuclear pleomorphism (HE stain, X200).

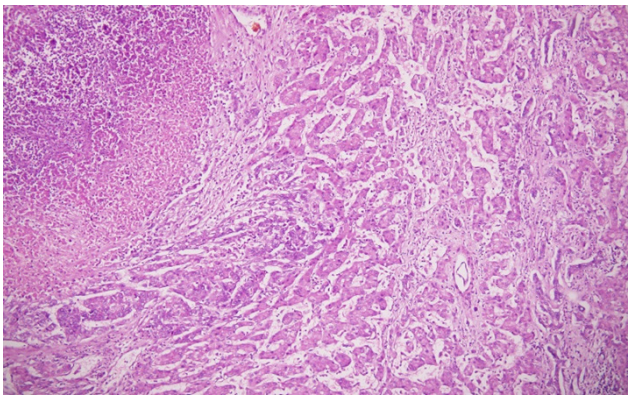

Figure 3: Fetal pattern with tumor cells arranged in thin trabeculae and adjacent areas show necrosis (HE stain, X100).

DISCUSSION

Hepatoblastoma is the most common primary malignant liver neoplasm in children.[1,2] Approximately 90% of the cases occur in patients under 5 years of age and two-thirds of the cases occur in the first 2 years of life.[6] Hepatoblastoma in adolescents and adults is very rare and is associated with poor prognosis compared to that in childhood. In adults, hepatoblastoma shows slight female preponderance. Review studies have shown that the age of adults with hepatoblastoma reported in the literature ranged from 17 to 18 years with a median age of 70 years for male and 27 years for female.[7]

Regarding etiology of hepatoblastoma, genetic studies have shown abnormalities in WNT/β-catenin signaling pathway in approximately 80% of cases.[8] Cytogenetic studies have revealed the involvement of chromosomal loci on 1q, 2(or 2q), 4q, 8(or 8q), and 20.[9] Furthermore, nuclear p53 accumulation indicates that p53 mutation is also involved in the molecular pathogenesis of the malignancy.[10] The pathogenetic pathway of this tumor is different in adults compared to children.[10,11]

Patients with hepatoblastoma present with nonspecific symptoms like abdominal pain, nausea, vomiting, fatigue, and fever. They usually have right upper quadrant pain, often in the presence of abdominal mass. Jaundice is uncommon in hepatoblastoma.

The initial diagnosis of hepatoblastoma is based on imaging. The role of MRI is more than that of ultrasonography and enhanced CT, in depicting tumor features like size, margins, and ratio with neighboring organs in order to the best surgical approach.[12]

Histopathology remains a gold standard for the diagnosis of hepatoblastoma. International Pediatric Liver Tumors Consensus Classification of Hepatoblastoma has classified hepatoblastoma as an epithelial type and mixed epithelial and mesenchymal type. Under epithelial type, some subtypes include fetal embryonal, small cell undifferentiated, cholangioblastic, macrotrabecular, and mixed epithelial.[13] These tumors are rarely composed of only one cell type, usually demonstrating combinations of epithelial, mesenchymal, undifferentiated, and other histologic components. The most common one is the embryonal pattern that resembles the liver at 6–8 weeks of gestation. A macrotrabecular growth pattern, similar to that typically seen in hepatocellular carcinoma, may be present in a minority of hepatoblastomas, accounting

for 05% of all cases. Cells within these macrotrabeculae may be fetal, embryonal, or pleomorphic and may be similar to those seen in hepatocellular carcinoma. Hepatoblastomas may contain undifferentiated small cells, sometimes coexpressing cytokeratin and vimentin, reflecting neither epithelial nor stromal differentiation. Small-cell-undifferentiated hepatoblastoma belongs to the clinically important group of hepatoblastomas that show low or normal serum AFP levels and have aggressive behavior. In addition to epithelial components, 20–30% of hepatoblastoma specimens also contain stromal derivatives, including spindle cells, osteoid, skeletal muscle, and cartilage, leading to the designation of 'mixed' hepatoblastomas.[13]

Immunohistochemistry is now being increasingly used in the diagnosis and classification of hepatoblastoma subtypes. A limited panel using at least alpha-fetoprotein, glypican 3, beta-catenin, glutamine synthetase, vimentin, cytokeratin (pankeratin), Hep-Par1, and INI1 are presently the most useful in this setting.[13]

Our case underwent complete surgical resection and is under chemotherapy. She has already received her two cycles of chemotherapy and is currently asymptomatic. Surgical resection is the mainstay of treatment for patients and a margin negative reaction is associated with better survival rates. Chemotherapy has been proven effective in the both adjuvant and neoadjuvant treatment and can cause tumour shrinkage.[14] However, hepatoblastoma presents a poor prognosis in the adult due to its metastatic behavior or the local aggressiveness.[4]

CONCLUSIONS

With the low incidence and non-specific initial symptoms, hepatoblastoma in adults presents a diagnostic challenge, demanding a high index of suspicion and thorough evaluation. Early detection, diagnosis, and treatment can improve the prognosis. As the disease is rare and can morphologically masquerade other malignant liver tumors primary or secondary, the pathologist needs to be aware of this rare entity in adults to make the correct diagnosis and benefit the patient.

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