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ISOLATED CEREBRAL METASTASIS OF HEPATOBLASTOMA IN A 3-YEAR OLD CHILD

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ABSTRACT

Hepatoblastoma (HB) is a rare tumor of infancy and childhood manifesting with hepatomegaly and abdominal distention. It is seen with increased frequency in association with congenital hemihypertrophy and renal anomalies. Metastases to other organs, including the lungs, lymph nodes and even ovaries is well known. But cerebral metastasis of this tumor, especially in the absence of metastasis to other organs, has not yet been reported. In this paper a 3-year old male child with congenital hemihypertrophy and isolated metastasis of HB to the brain is presented.

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INTRODUCTION

HB is a rare primary hepatic tumor of infancy and childhood.[3] The mean age at diagnosis is one year; most cases occur before 3 years of age. It is more common in males, and it may be associated with congenital anomalies, including congenital hemihypertrophy and renal anomalies.[4,7]

Most patients present with an abdominal mass or abdominal enlargement. Leukocytosis, thrombocytosis and mild anemia are also often noted.[4] An interesting sign is osteoporosis and pathologic fractures that reverse with tumor excision.[2] In 66% of patients, serum alpha-feto-protein (α-FP) is elevated,[4] and this is a useful marker for diagnosis, treatment response and follow-up of HB patients.[1] This tumor more often occurs in the right lobe of the liver, but may extend to involve both lobes.[7] Imaging studies show hepatomegaly, and calcification in 30% of cases. Lung metastasis occurs in 10-20% of cases, and lymph nodes are the second most common area.[4-6] Although cerebral metastasis of HB has been described, isolated metastasis of HB to the brain in the absence of tumor spread to other organs has not yet been reported.

CASE REPORT

A 3-year old boy with congenital hemihypertrophy presented with headache, vomiting, progressive decline in level of consciousnes and eventually coma. Neurologic examination showed central type facial paralysis, papilledema and a bilateral Babinski sign. Brain CT scan revealed a very large tumor in the right parieto-occipital area with nonhomogenous density, marked shift of the midline and ventricular dilatation (Fig. 1). The patient underwent craniotomy and the tumor was resected. Histopathologic examination of the specimen showed a tumor with large areas of necrosis, foci of undifferentiated mesenchymal tissue, osteoid formation, and epithelial portions highly reminiscent of hepatic trabeculae. Even accumulation of bile could be observed in some epithelial cells. A diagnosis of metastatic HB was given and liver imaging studies were requested. Ultrasonographic examination of the liver showed a large mass in the right lobe. Serum α-FP level was elevated up to 655 ng/mL (normal < 20 ng/mL) at this time. An open liver biopsy was performed and this confirmed the diagnosis of HB. The patient died soon afterwards.

Upon autopsy the right extremities were clearly larger than their left counterparts. A large 11-cm mass was found in the right lobe of the liver in the posterior part (Fig. 2). It was relatively well-circumscribed, soft and with extensive necrosis in central portions. Examination of paired internal organs such as lungs and kidneys showed asymmetry and enlargement of right side organs

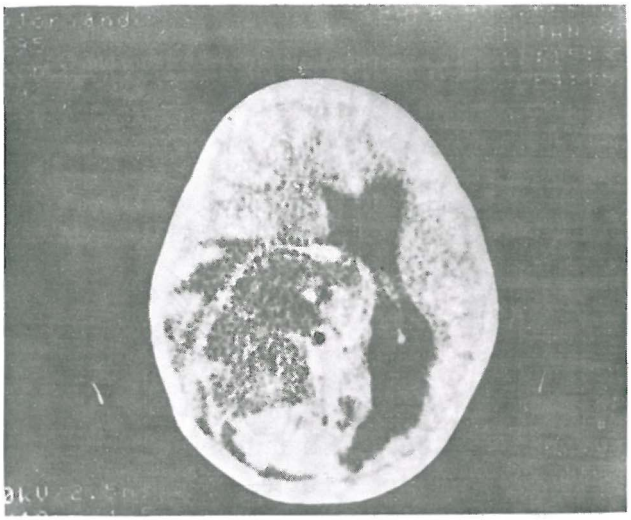

Fig. 1. Brain CT scan shows right parieto-occipital tumor.

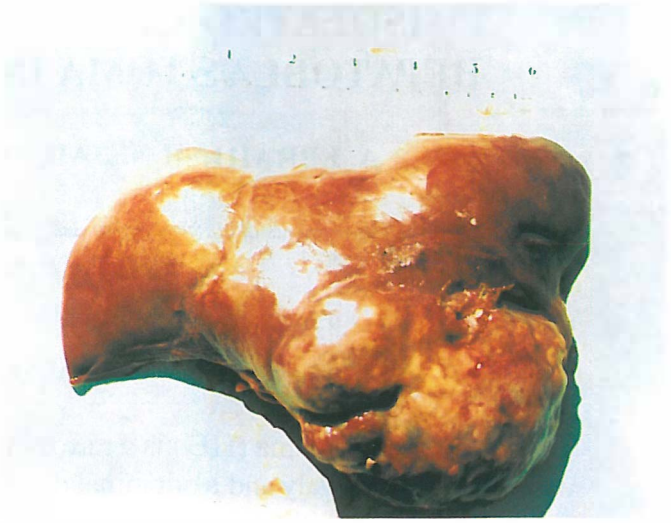

Fig. 2. Autopsy shows large hepatic tumor.

in comparison to the left organs. Histopathologic examination of liver hilar lymph nodes, lesser curvature nodes, celiac and para-aortic nodes, and pulmonary and intrathoracic lymph nodes did not show any metastatic deposit.

DISCUSSION

Congenital hemihypertrophy is a disease that is known to be associated with increased risk of tumor development, mostly Wilm's tumor. Similarly, HB occurs with increased frequency in these patients.[9] HB invades veins, and the lung is the most common site of tumor spread. Lymph nodes, adrenals, bones, ovaries and the brain are other sites for metastasis of HB.[5,6,8] However to our knowledge, isolated metastasis of HB to the brain has not been described.

The mainstay of treatment for HB is complete surgical resection of the primary tumor. HB is radiosensitive and chemosensitive; a regimen of vincristin, cisplatin and adriamycin can induce complete remission in many cases. Prognosis depends on the completeness of tumor resection.[1] When the tumor is completely excised, 3-year survival is 90%, but this figure drops to 10-20% in cases with distant metastases.

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