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Letter to the Editor

Multiple hepatic epithelioid hemangioendothelioma in an adolescent girl mimicking liver metastasis

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Dear Editor,

Epithelioid hemangioendothelioma (EHE) is low-to-intermediate grade malignant neoplasm of soft tissue vascular tumor of endothelial origin. It was first described by Weiss and Enzinger (1) in 1982; and it may arise from liver, lung, brain, bone, spleen, peritoneum, and soft tissues. The incidence is estimated at less than one case per million in the population, there is a female predominance (2). Little information is available in the literature about disease characteristics in children. We would like to present an adolescent girl with hepatic epithelioid hemangioendothelioma (HEHE) mimicking liver metastasis.

A previously healthy 17-years-old female was referred to us with multiple metastatic lesions in the liver. She had experienced vomiting for four days. Her medical history was otherwise unremarkable. It was learned that abdominal ultrasonography examination had showed multiple liver lesions involving both lobes (Figure 1A), and she had undergone upper gastrointestinal endoscopy for a possible metastatic gastrointestinal cancer in an adult gastroenterology clinic in another hospital.

Her physical examination was normal. The patient's hemogram was normal. The results of liver function tests were within the normal range. The patient was negative for the tumor markers carcinogenic embryonic antigen, and alpha fetoprotein. Liver CT demonstrated multiple lesions; all were hypodense at portal venous phase acquisition (figure 1B). Percutaneous liver biopsy was performed under US guidance. Liver biopsy revealed the diagnosis of HEHE. Repeated CT and ultrasounds showed that stable of all hepatic lesions for six years. The reported patient have a baby, the pregnancy and post-partum period for two years were eventful.

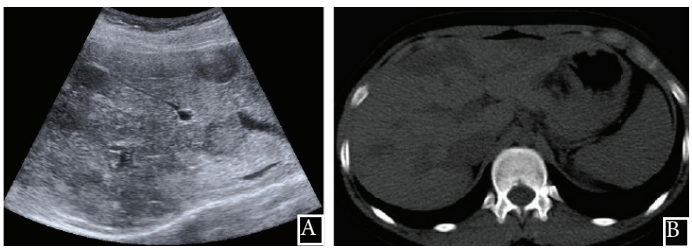

Figure 1 (A). Ultrasound showing multiple liver lesions **(B)**. Axial CT showing hypodense multiple lesions

The clinical course of EHE between benign hemangioma and angiosarcoma (2-4). Although HEHE follows behind embryonal sarcomas in frequency in the pediatric population; its remains the third most common reason for transplantation owing to unresectable liver tumors (2-4). Biological tests are usually normal and may elevated liver enzymes. Tumor markers are always normal. HEHE is treated with surgical resection or liver transplantation. Other treatments have been reported such as chemotherapy, radiotherapy, hormone-therapy, and embolization. The progression of HEHE is unpredictable (2-5). The spontaneous complete regression of a malignant tumor is very rare (6). Prolonged survival without therapy in patients with HEHE, beyond year, has been observed in few adults (7). Slow progression and partial spontaneous regression might occur, especially in asymptomatic patients. Kitaichi et al. (7) reported partial spontaneous regression of pulmonary EHE in three from 21 patients. Otrock et al. (6) reported complete regression of the HEHE in one patient.

As a conclusion, our experience underlines that; imaging studies of HEHE may present a diagnostic challenge. Thus a high index of suspicion is required for diagnosis. The course of a disease is uncertain, and there is no accepted treatment strategy.

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Conflicts of interest, The authors of this paper have no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

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