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PHOTO QUIZ

An uncommon cause of a giant abdominal mass

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Introduction

A 48-year-old female presents to our service for complains of vague intermittent right upper quadrant pain, she denied fever, night diaphoresis, and shortness of breath, nausea, vomiting or other symptom. Medical history was positive for a previous hepatectomy 17 years ago, and past tobacco smoking. She has no family history of liver disease or liver cancer. Physical examination revealed an abdominal mass located in the right hemiabdomen, not painful to palpation and not pulsating. Laboratory results were not significant and vital signs were within normal range. Magnetic resonance imaging (MRI) demonstrated a marked enlarged liver (Figure 1A and B), with a craniocaudal length of right lobe of 34 cm, extending down to the iliac fossa.

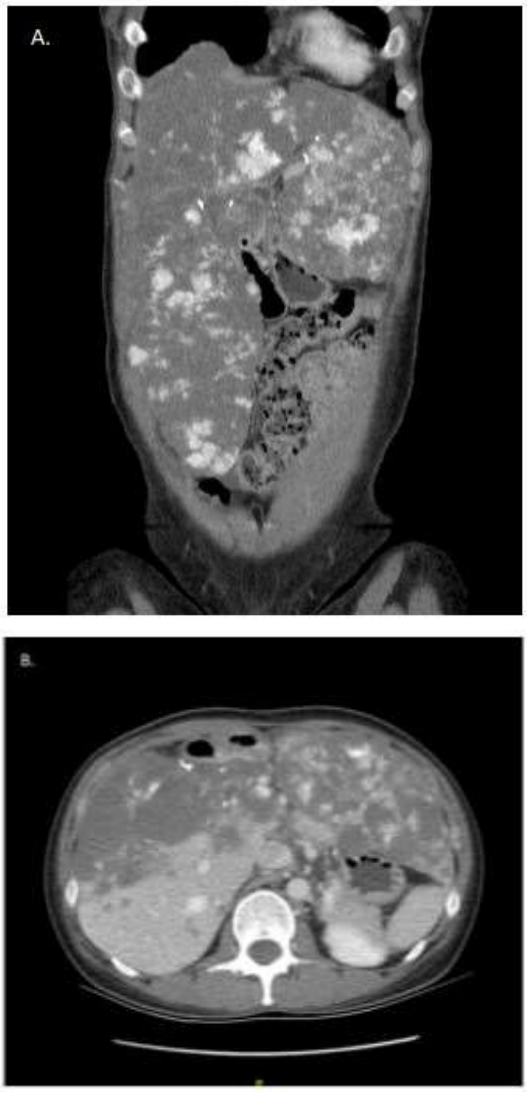

Figure 1. Contrast MRI of the abdomen in coronal plane (A) and axial plane (B) showing diffuse liver hemangiomas, replacing most of the normal parenchyma.

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What is the most likely diagnosis?

Diffuse hepatic hemangiomatosis (DHH). Hemangiomas is visible throughout the liver, additionally, there is a mass which is largest and most fluent in the right anterior and left medial segments and in segment three, replacing the entire parenchyma, possible consistent with a giant hemangioma.

What would be the best management?

The plan was determined to be a liver transplantation since the patient is symptomatic and because the diffuse hemangiomatosis is replacing the majority of the normal liver parenchyma.

Discussion

Hepatic hemangiomas (HH) are the most common benign liver tumors; they usually have an asymptomatic course and are found incidentally (1, 2). Adult women are more frequently affected, they are more commonly diagnosed toward the fourth and fifth decade of life (2, 3). They tend to be nodular solitary lesions and are commonly located in the right lobe of the liver (4). Hepatic hemangiomatosis and HH are different entities that share the same histology (5), however, both lesions coexist in a single patient 44 to 47% of times (6-9). Hepatic hemangiomatosis can be further subdivide into nodular or diffuse (DHH) (5). DHH is more common in neonates, with possible extrahepatic multi-organ affectation and a high mortality rate (10-12). The etiology of DHH remains unknown; it is a rare diagnosis in adults (13, 14) with only few cases published.

HH usually do not cause symptoms, abnormal findings in physical examination or liver blood tests and are frequently found incidentally while performing imaging studies for other reasons. Notwithstanding, some HH will exhibit symptoms, the most common being right upper quadrant abdominal pain (15) thought to appear due to stretching of the Glisson's capsule (1). Likewise, the main symptoms DHH of are abdominal pain and abdominal distention (1, 16-18). Laboratory panel results are usually normal or only with a mild elevation of ALT, AST, ALP and GGT (6). As these symptoms can be elicited by a wide variety of other diagnoses, it is highly important to rule out other possible etiologies in patients with HH or DHH that present with abdominal pain. Interestingly, akin to HH and other hepatic tumors, DHH size can fluctuate with estrogen therapy (19) and metoclopramide (20).

Possible complications of HH or DHH include bleeding from rupture (spontaneous or traumatic) (21) and Kasabach-Merrit syndrome, an infrequent but life-threatening coagulation disorder, characterized by disseminated intravascular coagulation, thrombocytopenia, and systemic bleeding (22, 23). The most common cause of death in patients with DHH is liver failure (6).

Imaging studies are key for the diagnosis of DHH; ultrasound (US) shows hepatomegaly and multiple hypoechoic nodules (6); computed tomography (CT) images display low-density nodules (6, 24), a with peripheral rim enhancement on late arterial phase sometimes is visible (5). MRI shows liver nodules with hypointensity on T1-weighted and hyperintensity on T2-weighted sequences (5).

Treatment is indicated for symptomatic patients or in case of complications. Surgery, either open or laparoscopically, remains the most common treatment for HH (23, 25). Recurrence is rare but has been reported with HH and DHH (26, 27). Furthermore, liver transplantation is indicated for patients with HH or DHH who are ineligible for resection (28-30).

Conclusion

In conclusion, although HH and DHH share a histologic pattern, they are different entities. They represent an unusual indication for liver transplantation. As patients usually present with unspecific symptoms, physicians need to be familiar with their aspect on imaging studies to make accurate diagnoses.

Conflict of interests

The authors declare that they have no conflict of interest.

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