Primary Hepatic Epithelioid Hemangioendothelioma Masquerading as a Hepatic Abscess With Infective Picture: A Case Report

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Abstract
Hepatic endothelioid hemangioendothelioma (EHE) commonly presents with multiorgan involvement with locally aggressive behavior. In most cases, it presents with right upper quadrant abdominal pain, hepatomegaly, and weight loss with metastasis commonly to the lung. We present a 33-year-old woman with hepatic EHE with an initial presentation mimicking hepatic abscess and imaging findings misleading for metastatic liver lesions. It was confirmed on pathology with immunohistochemistry, but the patient could not survive due to her late presentation and the presence of metastatic lesions in the lung.

Introduction
Epithelioid hemangioendothelioma (EHE) is an unusual endothelial or preendothelial cell vascular tumor formed by epithelioid and histiocytic appearing cells in a mucoid to fibrotic stroma [1]. It has been reported with an incidence of fewer than two cases per 1 million [2]. In the 2020 World Health classification of soft tissue tumors, epithelioid hemangioendothelioma was classified as a malignant vascular tumor because of its 15% chance of metastasis, especially when located in the lungs and the pleura [3]. Patients might present with symptoms such as weight loss, liver enlargement, and pain in the right upper quadrant of the abdomen [4]. The diagnosis of hepatic EHE is confirmed with pathological examination with immunohistochemistry.

We present a rare hepatic epithelioid hemangioendothelioma in a female patient which initially resembled a hepatic abscess.

Case Presentation
A 33-year-old woman who was previously healthy was admitted to the hospital with complaints of cough, fever, chills, and right-sided abdominal pain for three months. The abdominal pain was not related to food intake. No change in stool color and no change in bowel habits were noted. The patient denied any similar episodes in the past. Vital signs were as follows: temperature: 37.5 °C, heart rate:100 beats per minute, respiratory rate: 16 breaths per minute, and blood pressure: 112/70 mmHg. On general examination, the patient was conscious and oriented. Abdominal examination elicited right upper quadrant tenderness. Other systemic examinations were within normal limits. Laboratory results showed high inflammatory markers, eosinophilia, microcytic anemia, and elevated liver function tests.

A chest radiograph showed no evidence of clear infiltration. An ultrasound abdomen was done, which showed an enlarged liver with multiple calcifications. An MRI abdomen was done which showed an enlarged liver and the right lobe demonstrated heterogeneous bright T2 weighted signal intensity with mild diffusion restriction associated with heterogeneous parenchymal enhancement on the postcontrast sequences. There were multiple space-occupying lesions, the largest seen in segment V of the liver with ill-defined margins, mildly bright on T2 weighted images showing predominant peripheral enhancement suggestive of an early hepatic abscess. Smaller satellite lesions were also seen in the right lobe. Abnormal low T2 weighted signal intensity changes are seen in an infiltrative manner in the right lobe of the liver superior to the abscess with focal signal void areas (Figure 1). These findings were suggestive of hepatic abscesses in the right lobe with surrounding likely granulomatous hepatitis.
Entamoeba serology came positive and the patient was started on intravenous metronidazole, paromomycin, and ceftriaxone. A CT thorax, abdomen, and pelvis were performed, and they found a large, ill-defined liver lesion that was non-enhancing and multiple satellite lesions in both lobes. Multiple chest nodules were seen bilaterally (Figure 2).
FIGURE 2: CT abdomen and pelvis with contrast, (A) axial and (B) coronal portal phase, showing enlarged liver with large ill-defined hypodense area in the right lobe (red arrows). (C) Axial lung window showing bilateral lung nodules.

Whole-body FDG PET (fluorodeoxyglucose positron emission tomography) CT showed intense diffuse uptake in the right lobe of the liver, suggestive of diffuse involvement with minimal involvement of the left lobe. Suspicious portocaval metastatic lymph nodes and lung nodes were also seen (Figure 3).

FIGURE 3: (A) Axial and (B) coronal PET CT showing intense diffuse uptake of FDG in the right lobe suggestive of diffuse involvement (yellow arrows).

Liver biopsy showed pleomorphic epithelioid cells, with occasional intracytoplasmic vacuoles, seen within a myxohyaline stroma. The tumor cells were diffusely positive for vascular markers, namely CD31, CD34, and ERG (Figure 4). Cytokeratins and liver-specific markers such as HepPar1 and arginase 1 were negative. The morphology and immunohistochemistry were in keeping with a diagnosis of epithelioid hemangioendothelioma. Bone marrow (BM) aspirate was hypercellular with trilineage hematopoiesis and prominently increased erythroid precursors with severe dyserythropoiesis. No increase in blasts was noted. Viral serology, fungal culture, and tuberculosis tests were negative. Hb electrophoresis revealed the B-thalassemia trait.
FIGURE 4: (A) Hepatic hemangioendothelioma composed of pleomorphic epithelioid cell (red arrow) with background myxohyaline stroma, (B) foci of tumor cells with prominent intracytoplasmic vacuoles (black arrow), (C) CD31 immunostaining highlighting the vascular nature of the lesional cells (blue arrow), and (D) ERG immunostaining demonstrating strong nuclear positivity (green arrow).

The case was discussed in the hepatobiliary multidisciplinary team and the advice was for the right hepatectomy by the hepatobiliary surgeon. The patient started deteriorating on day 7, had multiple episodes of vomiting, and complained of severe pain in her right upper quadrant. Repeated blood investigations revealed increased CRP, prolactin, leukocytes with eosinophils, and thrombocytopenia. The patient further went into acute liver failure and hepatic encephalopathy. She was started on IV meropenem, tazocin, and steroids. She was shifted to the medical intensive care unit following a loss of consciousness. A CT head revealed no intracranial pathology. She went into disseminated intravascular coagulation and, unfortunately, could not survive.

Discussion

According to the study by Mehrabi et al., the presentation of hepatic EHE is variable and can range from no symptoms at all to full-blown liver failure. Although a quarter of the patients were asymptomatic on presentation, the most common presenting symptoms for patients were weight loss, liver enlargement, and pain in the right upper quadrant of the abdomen. It was also noted that a significant proportion of the patients with hepatic EHE had some other organ involvement, with pulmonary metastasis leading the list [4]. Llueca et al. reported a patient presenting with a pelvic mass with liver metastatic lesions who was later found to have hepatic EHE [5], which shows how unpredictable the presentation of EHE can be.

EHE has a propensity to occur in the liver, lung, pleura, spleen, bone, brain, meninges, breast, stomach, lymph nodes, soft tissue, thyroid, prostate, and ovaries. It has been reported in more than 200 cases worldwide so far, and it most commonly occurs between the ages of 20 and 60 years old, with a predominance in females [2]. Hepatic EHE commonly presents with multilobar involvement with locally aggressive behavior. It is similar to angiosarcoma pathologically as a vascular origin tumor, however, with a relative improved prognosis. In most cases, it presents with right upper quadrant abdominal pain, hepatomegaly, and weight loss with metastasis commonly to the lung [4].

The main imaging features of hepatic EHE on magnetic resonance imaging (MRI) or computed tomography (CT) are the peripheral location of the nodules, the contraction of the capsule, and the tendency of multiple foci to merge [6,7]. It has been noted that the “lollipop sign” in which hypodense mass (the candy in the lollipop) on contrast with the termination of vascular stock (the stick) as it abuts the mass and the “target
Hepatic epithelioid hemangioendothelioma is a rare liver tumor with a variety of differential diagnoses, and early diagnosis and treatment significantly reduce mortality and morbidity. Appropriate evaluation includes imaging and biopsy. Treatment consists of surgical excision for localized disease and liver transplantation for extensive liver infiltration. Chemotherapy and ablation may also be considered for metastatic disease.

Conclusions

Hepatic epithelioid hemangioendothelioma is a rare liver tumor with a variety of differential diagnoses, and early diagnosis and treatment significantly reduce mortality and morbidity. Appropriate evaluation includes imaging and biopsy. Treatment consists of surgical excision for localized disease and liver transplantation for extensive liver infiltration. Chemotherapy and ablation may also be considered for metastatic disease.

Additional Information

Disclosures

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