Fellows' Corner

by Captain Roselle M. Hoffmaster, M.D., Captain Edward C. Bergen, M.D.
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CASE REPORT

A 23-year-old male presented with a two day history of painless jaundice. Past medical history was notable for a prior history of abdominal pain four years previously with mild elevations in AST and ALT. Evaluation with an EGD and colonoscopy were unremarkable at that time and an abdominal ultrasound (US) was negative for liver parenchymal abnormality. On presentation, the patient was found to have conjunctival icterus, hepatosplenomegaly, and erythematous macules on the right side of his neck. Lab evaluation was significant for a mixed pattern of elevated liver enzymes in a predominantly cholestatic pattern and decreased synthetic function: GGT 528 IU/L, AP 884 IU/L, AST 157 IU/L, ALT 138 IU/L, TB 10.4 mg/dL, DB 8.3 mg/dL, Albumin 3 g/dL, and INR 1.3. An abdominal US showed a heterogeneously enlarged liver and a small amount of ascites. Computerized tomography (CT) showed numerous sub-centimeter pulmonary nodules throughout all lung fields, portal hypertension, and multiple enhancing hepatic nodules. An US guided liver biopsy stained positive for the endothelial marker, factor VIII. Arterial phase liver CT and liver biopsy (100× hematoxylin and eosin stain) are shown below.

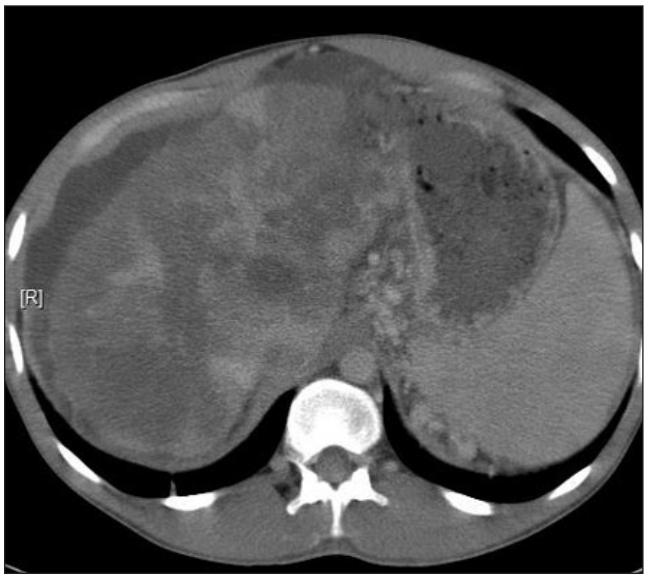

Figure 1.

Questions

1. What is the diagnosis?
2. How does this disease usually present?
3. What are the treatment options?

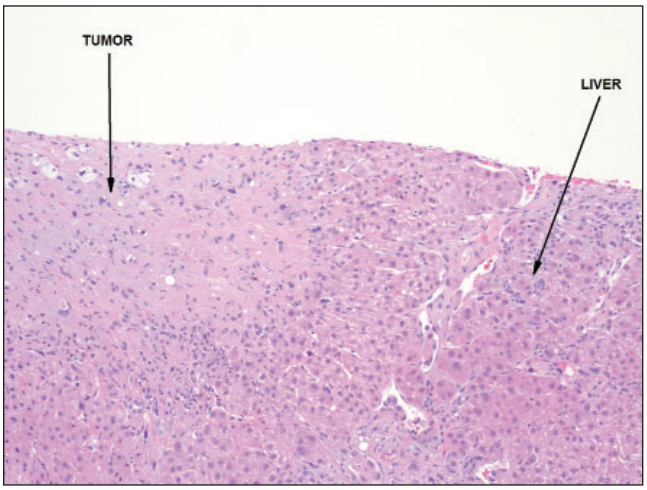

Figure 2.

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DISCUSSION

The patient was diagnosed with hepatic epithelioid hemangioendothelioma (HEH), based on characteristic pathology: tumor cells with intracytoplasmic lumens containing erythrocytes and positive staining for endothelial markers, including factor VIII. HEH is a rare tumor of vascular origin with approximately 300 known cases reported in the literature since its description in 1984 (1). There is a slight female predominance with a mean age at diagnosis of 46 (2). Early case series postulated a link with use of oral contraceptives, but no known etiology has been found.

Patients usually present incidentally or with nonspecific symptoms including right upper quadrant pain, weight loss, and fatigue (2). Less common presentations include jaundice, Budd-Chiari syndrome, or fulminant liver failure (3). Elevations in hepatic enzymes with a cholestatic pattern predominant and CT shows concentric nodules or diffuse involvement with delayed arterial phase enhancement, but the diagnosis is made via pathology with staining for endothelial markers.

The tumor exhibits behavior in between that of a benign hemangioma and an aggressive angiosarcoma. Without treatment, 5-year survival rates vary widely from 28%–67% (1,2). In one series, 27% of patients have metastatic disease at the time of diagnosis, most commonly to the lungs, spleen, bone, or abdomen (2). Even with metastatic disease, reports in the literature note prolonged survival and spontaneous regression. Treatment strategies have included partial hepatectomy, chemotherapy, radiation therapy, transarterial embolization, and liver transplant. For patients with unresectable disease, liver transplant currently offers the most promise with 5- year survival rates of 43%–76%, including patients with metastatic disease (4,5,6).

References

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