Metastatic Hepatic Epithelioid Hemangioendothelioma: A Case Report

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Introduction

- Epithelioid Hemangioendothelioma (EHE) is a very rare vascular tumor which tends to behave as a low-grade malignancy despite a high inclination for systemic involvement.
- Peaks in the 4th-5th decade with a slight predominance in females compared to males.

Case Information

History of Present Illness:
A previously healthy 47-year-old male, that initially presented to the emergency department with right upper quadrant pain, low grade fever, and appetite loss which had been present for 2 weeks.

Past Medical History:
No relevant prior medical history, non-smoker, 1-2 alcoholic beverages 2-3 nights per week, family history of skin cancer, ulcerative colitis, and lung cancer

Investigations:

- Initial presentation was concerning for cholecystitis therefore an US and CT abdomen and pelvis were performed which found a complex lesion in the right hepatic lobe measuring 6.5 X 5.3 cm.
- CT guided core needle biopsy was performed to characterize lesion which was significant for atypical cells seen within a somewhat loose myxoid background, occasionally showing cytoplasmic vacuoles.
- Immunohistochemistry revealed CD31 and CD34 positivity and a strong positive CAMTA-1 nuclear staining further confirming EHE.
- Additional CT chest demonstrated several small bilateral pulmonary nodules, the largest measuring 1.1 cm in the left upper lobe increasing concern for metastasis.

Images

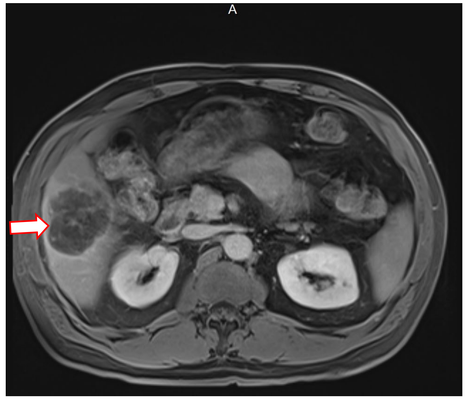

Figure 1: T2 Axial Vibe FS Delay 5min with lesion (arrow)

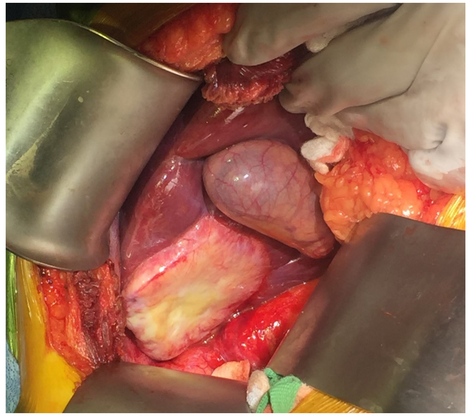

Figure 2: Intraoperative photo of mass to right liver

Case Information (cont'd)

Course:

- Underwent right total hepatectomy (seg 5, 6) with associated cholecystectomy and wedge biopsy of suspicious lesion in the right lateral abdominal wall parietal peritoneum overlying diaphragm.
- Liver path demonstrated a 6.5 cm liver mass consistent with hepatic EHE with negative surgical resection margins and background liver with moderate steatosis (30-40%).
- Immunohistochemistry was positive for ERG, CD34 and TFE3.
- The specimen demonstrated moderate nuclear pleomorphism, 1 mitosis per 10 HPF, and showed patchy necrosis (approximately 15% of tumor).
- Diaphragm path revealed metastatic EHE with positive immunohistochemical staining for ERG in the tumor cells.
- Given Dx of hepatic EHE with stage IV mets, Oncology planned for CT chest/abdomen/pelvis as part of standard surveillance with additional imaging only as indicated.
- Given that his pulmonary nodules were small and indeterminate at the time, close observation was favored with follow-up and repeat imaging q3m X 1yr then q4m X 1yr then q6mo presuming stable disease.
- At 15 months s/p surgery, imaging continued to reveal unchanged bilateral pulmonary nodules, stable to slightly small seroma, unchanged left renal lesions, unchanged omental infiltration and nodularity particularly in the RUQ suspicious for sarcomatosis which will be monitored.

Implications/Discussion

- Arises anywhere throughout the body but tends to mostly involve the liver, lungs, and bone with >50% of patients presenting with mets.
- Clinical presentation varies widely from asymptomatic to symptomatic with the most common symptoms being pain (40%), a palpable mass (6%-24%) and weight loss (9%).
- Etiology of the disease is still not fully understood, but a few possible associated factors including vinyl-chloride, oral contraceptive use, viral hepatitis and trauma have been reported.
- Histologically, the tumor has been mistaken for metastatic carcinoma, angiosarcoma, hepatocellular carcinoma, and cholangiocarcinoma.
- Stains positive for vascular markers, such as factor VIII–related antigen, CD31, and CD34, and negative for cytokeratins.
- CAMTA1- WWTR1 fusion product is the most identified genetic abnormality with this tumor but the pathway of how this fusion leads to oncogenesis has yet to be elucidated.
- Biopsy with histological analysis and various immunohistochemistry staining are done for definitive diagnosis.
- Currently no consensus in treatment protocol, techniques including liver transplantation, liver resection, chemo and/or radiation therapy, and surveillance have all been used with varying outcomes.
- Chemo, typically employing traditional chemotherapies used for other soft tissue sarcomas (e.g. anthracycline-based regimens), or anti-angiogenic approaches (e.g. pazopanib), would be important if he develops significant disease progression.

References

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- Errani, C., Zhang, L., Sung, Y. S., Hajdu, M., Singer, S., Maki, R. G., ... & Antonescu, C. R. (2011). A novel WWTR1-CAMTA1 gene fusion is a consistent abnormality in epithelioid hemangioendothelioma of different anatomic sites. *Genes, Chromosomes and Cancer*, *50*(8), 644-653.