Mediastinal Epithelioid Hemangioendothelioma with Osseous Metaplasia - A Case Report on an Ultra-Rare Sarcoma

Jamie Whisler[1] DO; Austin Forbes[1] DO; Daniel Cuadrado[1] MD
[1]Saint Joseph Medical Center, Tacoma, WA, USA

Background

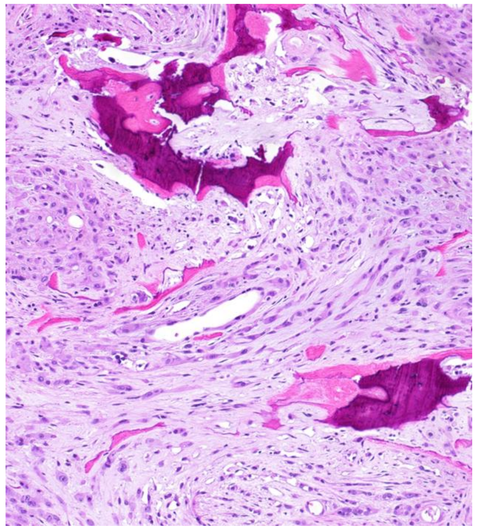
Epithelioid hemangioendothelioma (EHE) is a malignant vascular tumor that was first described in 1972 and not named until 1982. Literature currently includes 248 cases total with a 4:1 female preponderance and median age of 36 at diagnosis. The majority of the tumors were found in the liver, followed by cases in the lung, soft tissue, and then bone; most patients presented with multiple tumors and multiple involved organs. Only 12 cases of mediastinal instances were found and often thought to arise from the superior vena cava or brachiocephalic vein. Clinically this condition ranges in behavior from benign hemangioma to high-grade angiosarcoma. Median survival is 4.6 years with a range from 6 months to 24 years; the majority of mortality occurs from respiratory failure due to tumor burden in the lungs.

Case Presentation:

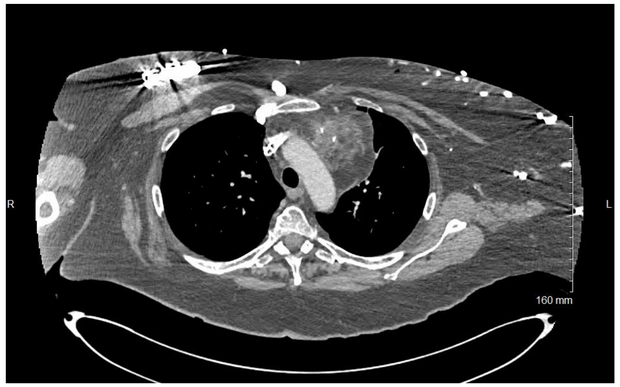
A 68 year-old female presented with an anterior mediastinal mass found incidentally during a CT scan done as work-up for pacemaker placement. The mass was noted to be 6.1 x 4.1 x 5.0 cm of irregular lobulated fat/soft tissue mass with interspersed calcifications. Imaging differential included thymoma, thymic carcinoma, thymic rebound, metastasis, thymolipoma, or germ cell tumor. CT-guided biopsy demonstrated extramedullary hematopoiesis. Surgery involved en bloc resection of the left phrenic and left vagus nerve due to frank involvement and partial invasion of the innominate vein was noted but dissection robotically was still feasible. Resection margins were negative with one negative lymph node. Her postoperative course was significant for elevation of the left hemidiaphragm and persistent hoarseness without any shortness of breath. She has completed a vocal cord injection with ENT with some improvement in her symptoms. Her pathology has undergone expert review with additional outside consultation. She was referred to medical oncology at a sarcoma center who recommended interval surveillance every 3-6 months with FDG-PET/CT for 2-3 years as currently little benefit has been shown with adjuvant therapy.



6.1 x 4.1 x 5.0 cm soft tissue mass with calcifications, found incidentally during work-up for pacemaker.



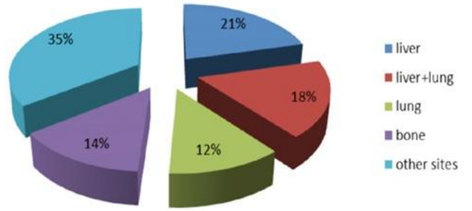
Osseous metaplasia with destruction of pre-existent bony trabeculae.



Mediastinal mass abutting the aorta, involving the left phrenic and left vagus nerve.

Discussion

EHE occurs in less than one in one million and therefore very little is known about the disease. Because the course may be incredibly indolent or aggressive, treatment is difficult to decide but typically involves surgery and potentially chemotherapy and/or radiation. The majority of mediastinal patients were asymptomatic and incidentally discovered, with diagnosis not occurring until postoperative pathology, such as in the case above. Often during surgery, the tumor is seen to be invading a vessel, but postoperative insight signifies the tumor may have developed from the vessel. While rare, keeping this on the differential may benefit patients with otherwise little known disease.



Most common locations of EHE upon presentation.

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