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Pleural Epithelioid Hemangioendothelioma

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A 51-year-old female presented to the emergency department complaining of left-sided, nonpleuritic chest pain and a 10-lb. weight loss over the previous 2 months. She was a current smoker with a 20 pack-year history. A PA/lateral chest radiograph and chest computed tomogram (CT) demonstrated a large loculated effusion with pleural thickening (Fig. 1). There were no lung nodules or pathologic mediastinal or hilar lymphadenopathy.

A diagnostic thoracentesis was performed, and serosanguineous pleural fluid was obtained. Cytology was negative. For further diagnostic sampling, she underwent a thoracotomy with pleural fluid drainage and pleural biopsy, which demonstrated sheets of atypical epithelioid cells displaying prominent intra-alveolar growth (Fig. 2). The cells stained positive for CD31 and CD34. Stains for pan-cytokeratin, CK5-6, p63, calretinin, S-100, Melan-A, TTF-1, CD68, and EMA were negative. She was given a diagnosis of pleural epithelioid hemangioendothelioma.

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In the multidisciplinary oncology conference, it was felt she was not a candidate for surgical resection given the extensive nature of the tumor. She was offered chemotherapy with doxorubicin. Less than 4 months after her initial presentation, the patient was admitted to the hospital with a bowel obstruction. Repeat imaging demonstrated further extension of tumor in the left hemithorax with associated volume loss, an enlarging right-sided pleural effusion, evidence of liver and spleen metastasis, and new-onset ascites. Her condition continued to deteriorate with the onset of respiratory failure and she died several days later.

Discussion

Epithelioid hemangioendothelioma (EHE) is a malignancy of vascular endothelial origin often involving bone, liver, soft tissues, and lung. Pleural EHE is rare, usually affects males, and is associated with a grim prognosis with disseminated disease [1–3]. Pleural involvement in females is even more unusual. In a recent review of 22 patients with pleural EHE, 82% were men and the average survival was only 10 months [1]. Radiographically, pleural EHE is nonspecific and can be easily mistaken for pleural infections or malignant mesothelioma. Chest CT usually demonstrates a pleural effusion with localized or diffuse nodular pleural thickening similar to our case [3–5].

Diagnosis is confirmed by pathologic examination of specimen obtained by surgical lung biopsy. On immunohistochemistry, EHEs show the typical phenotype of endothelial cells with reactivity to vascular-specific markers, including Factor VIII-related antigen, CD31, CD34, and Fli-1 [5]. The identification of Weibel-Palade bodies and pinocytotic vesicles on electron microscopy can also be useful in the diagnosis of EHE but is

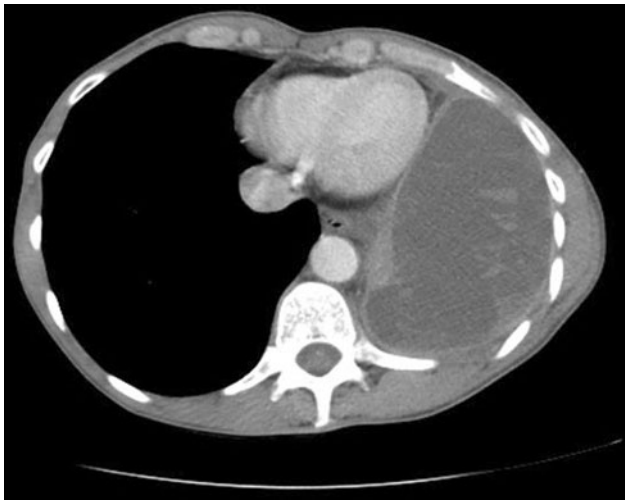

Fig. 1 Chest CT demonstrating a large, loculated pleural effusion with diffuse pleural thickening

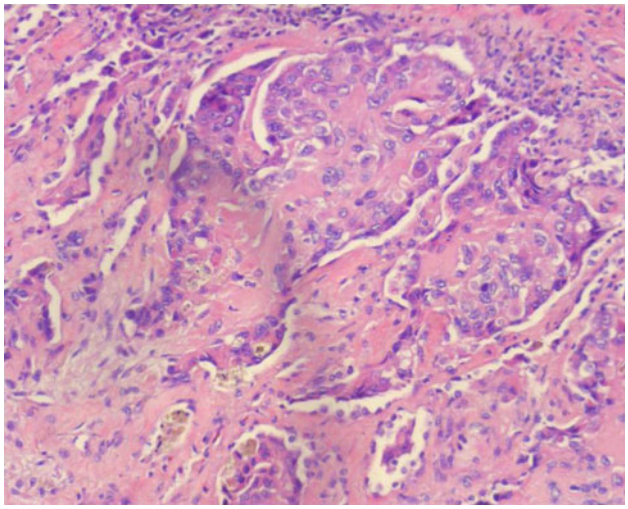

Fig. 2 Hematoxylin and eosin stain showing atypical epithelioid cells with cytoplasmic vacuoles

usually not required if immunohistochemistry confirms the diagnosis [5].

A standard therapeutic approach for pleural EHE has not been established. Surgical resection has been cited in the literature as the treatment of choice if the disease is limited [1, 3]. Unfortunately, the extensive nature typical of pleural EHE often precludes the utility of surgery, which was the case in our patient. A variety of chemotherapy regimens, including carboplatin, etoposide, paclitaxel, gemcitabine, doxorubicin, and interferon, have been tried with often limited or no benefit. There are currently no known effective or recommended chemotherapy regimens for pleural EHE.

Conflict of interest The authors have no conflicts of interest to disclose.

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