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Case Report

Mediastinal epithelioid hemangioendothelioma—Mimicker of mature teratoma

*Alexandre Semionov**, *Mutaz Khairo*

McGill University Health Centre, Department of Diagnostic Radiology, Montreal General Hospital, 1650 Cedar Avenue, Montreal, QC H3G 1A4, Canada

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ABSTRACT

We report a case of an adult man who underwent surgical resection of a presumed anterior mediastinal teratoma, which upon histopathologic assessment was diagnosed as an epithelioid hemangioendothelioma (EHE). EHE is a rare vascular tumor of variable, but usually low malignant potential. Its occurrence in mediastinum is estimated at 1 in a million. As EHE often contains macroscopic fat and bone, it can be indistinguishable on imaging from mediastinal mature teratoma. Therefore, EHE should be included in differential diagnosis of fat and bone-containing mediastinal masses.

Introduction

This is a case of mediastinal epithelioid hemangioendothelioma, a rare low-malignant potential tumor, misdiagnosed on CT imaging as a mature teratoma.

Case presentation

A 52-year-old man was referred to our institution for resection of an incidentally discovered anterior mediastinal mass. The lesion was discovered on CT chest performed for evaluation of vague chest discomfort experienced by the patient for roughly a year. A 3.6 by 3.2 by 3.8 cm well-defined lesion in the anterior mediastinum was originally discovered on CT chest performed for unrelated reasons. The lesion was predominantly of fat attenuation with central calcifications suggestive of bone (Fig. 1), and was originally diagnosed as a mature teratoma based on its imaging characteristics. The patient was further evaluated with a PET CT, which showed mild FDG-activity in the anterior mediastinal lesion and no other hypermetabolic foci to suggest metastases (Fig. 2).

The lesion was surgically excised. Involvement of portions of the left brachiocephalic vein, left internal mammary artery and left phrenic nerve were discovered at the time of surgery, necessitating partial resection of these structures.

Histopathologic analysis of the lesion demonstrated inflamed adipose tissue, spindle cell proliferation, and occasional bone and epithelial clusters. The spindle cells stained with ERG, CD31 and CD34, in keeping with the diagnosis of EHE. The resected tissue showed tumor-free margins.

* Corresponding author.
E-mail address: alexandre.semionov@mail.mcgill.ca (A. Semionov).
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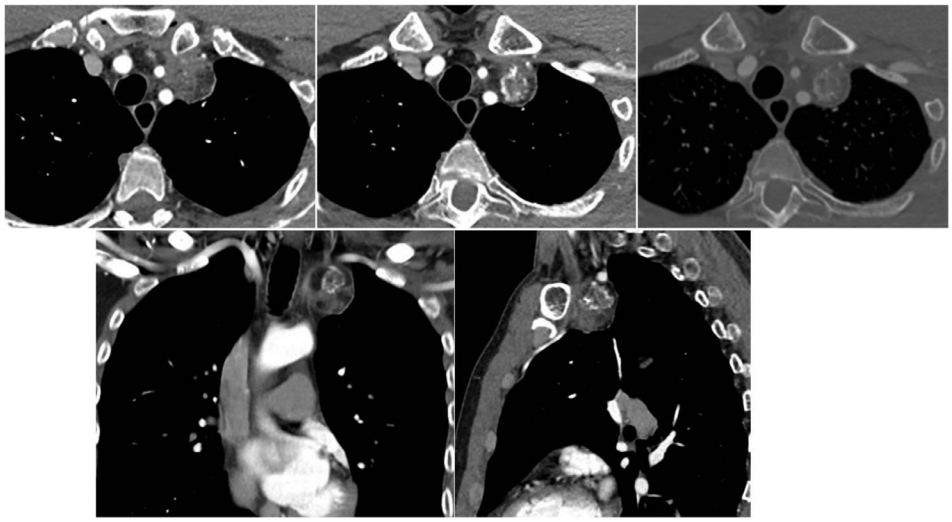

Fig. 1 – Contrast enhanced axial CT of the chest, and coronal and sagittal reformations demonstrate a well-defined left anterior mediastinal mass containing fat, soft tissue of muscle attenuation, and irregular calcifications—imaging characteristics suggestive of a mature teratoma.

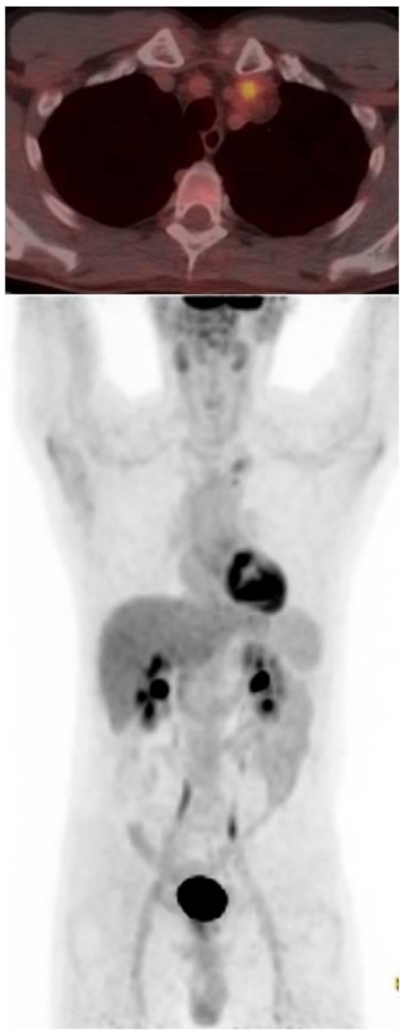

Fig. 2 – PET CT images demonstrate mild uptake in the left anterior mediastinal lesion. There are no other hypermetabolic lesions to suggest metastases.

Discussion

Hemangioendothelioma is a rare vascular neoplasm, classified by WHO as a low-grade angiosarcoma, which may arise in soft tissue, bone, lungs, liver, and occasionally in central nervous system, lymph nodes and breast [1]. Mediastinal EHEs are exceedingly rare [2].

The largest series describing mediastinal EHE published to date is by Suster et al, who report 12 such cases [3]. In their series, EHE occurrence showed male predominance (9:3); mean patients' age of 49 years (19-62 years range); presence of symptoms related to mass effect on adjacent mediastinal structures in 7 patients. All tumors were surgically removed. The excised tumors ranged in size from 4.5 to 13.5 cm; 7 lesions were well-circumscribed, and 5 tumors were locally infiltrative. Metaplastic bone formation and osteoclast-type giant cells were observed in 5 cases on histopathology. Only 1 patient from this series had a recurrence on follow-up.

A number of additional case reports of mediastinal EHE have been published, and frequently describe tumors involving or arising from mediastinal venous structures: superior vena cava, azygos, and brachiocephalic veins [4,5].

In all the published cases the diagnosis of EHE was made postoperatively following surgical removable of what was believed to be mature teratomas.

In conclusion, imaging features of mediastinal EHE are often indistinguishable from mature teratoma, and its correct diagnosis requires histopathologic analysis.

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