Clinical Case Report

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Angiomyofibroblastoma of the mediastinum

A case report and literature review

Jianguo Zhu, MD, PhD*, Sheng Su, MD, Haige Li, MD

Abstract

Introduction: Angiomyofibroblastoma (AMFB) is an extremely rare disease. It commonly occurs in middle-aged females and mainly involves the vulvovaginal region. Pathological examination plays an important role in differentiating from other tumors. But far less published literature focus on the imaging characteristics of AMFB.

Clinical findings/diagnoses: We reported a case of AMFB in a 73-year-old male, involving the mediastinum with computed tomography (CT) imaging and pathologic findings. Preoperative unenhanced CT scan of chest revealed a $6.9 \times 7.4 \times 9.3$ cm mass with equal density, located in the posterior mediastinum. On contrast-enhanced CT images, the tumor presented moderate, heterogeneous enhancement. Due to the unclear interface between the tumor and adjacent tissues, this patient underwent operative partial resection of the tumor. The pathologic diagnosis was AMFB.

Conclusions: This tumor represents a further example of unusual location different from conventional AMFB. This case adds to the experience with AMFB by summarizing its characteristics, and also reviewing the literature.

Abbreviations: AAM = aggressive angiomyxoma, AMFB = angiomyofibroblastoma, CA125 = serum carbohydrate antigen 125, CA724 = serum carbohydrate antigen 724, CEA = carcinoembryonic antigen, CK19 = cytokeratin 19, CT = computed tomography, HU = Hounsfield unit, NSE = neuron-specific enolase, PSA = serum prostate-specific antigen, SFT = solitary fibrous tumor.

Keywords: angiomyofibroblastoma, CT, mediastinum, pathology

1. Introduction

Angiomyofibroblastoma (AMFB) is an uncommon mesenchymal tumor that most predominantly occurs in the vulvovaginal area of middle-aged females, such as the vulva, perineum, and vagina. Occasional cases have been reported to arise in the scrotum and the inguinal region in males.[1] AMFB is a slow-growing soft-tissue neoplasm characterized by benign biological behavior. To our knowledge, only 1 case of recurrence has been reported in the English literature.[2] Histological findings of AMFB show alternating hypo and hypercellular areas mixed with thin-walled blood vessels with a perivascular concentration of stromal cells.[3] In this study, we report the first case of AMFB of the mediastinum occurring in a 73-year-old man and describe the computed tomography (CT) features. The aim of the current study was to better our understanding with AMFB by summarizing its characteristics, and also reviewing the literature. This study was approved by the institutional review board at the Second Affiliated Hospital of Nanjing Medical University in China, with informed patient consent.

2. Case report

A 73-year-old male was admitted to our hospital because of dysphagia for 1 month. His laboratory data such as complete blood cell count and creatinine showed no significant abnormalities. Tumor markers (serum carbohydrate antigen 125 [CA 125], serum carbohydrate antigen 724 [CA724], cytokeratin 19 [CK19], carcinoembryonic antigen [CEA], serum prostate-specific antigen [PSA], and neuron-specific enolase [NSE]) were within normal limit. Preoperative unenhanced CT scan of chest revealed a $6.9 \times 7.4 \times 9.3$ cm oval mass with equal density (mean CT value 25 HU), located in the posterior mediastinum (Fig. 1). On contrast-enhanced CT images, the tumor presented moderate, heterogeneous enhancement. The mean CT values were 40 HU (range 34–52 HU) in arterial phase and 64 HU (range 40–71 HU) in venous phase (Fig. 1). The tumor had unclear interface with adjacent tissues causing pressure on the esophagus, surrounding the descending aorta and causing localized atelectasis (Fig. 2). Due to the unclear boundaries between the tumor and descending aorta and pericardium, forcible separation of the tumor may actually raise the risk of potentially deadly complications such as massive bleeding and pericardial rupture. So, this patient underwent operative partial resection of the tumor to relieve the oppression on the esophagus. Microscopically, the tumor was composed of small cells embedded in a prominent myxoid/edematous stroma with hypocellular and hypercellular areas. The abundant vessels of various wall thicknesses were distributed in the collagenous stroma. The stroma consisted of spindle stromal cells with eosinophilic cytoplasm (Fig. 3). With immune stains, the tumor cells were positive for vimentin, desmin, and CD34, and negative for S-100. The proliferation index, expressed as a

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The authors report no conflicts of interest.

Department of Radiology, the Second Affiliated Hospital of Nanjing Medical University, Nanjing, China.

** Correspondence: Jianguo Zhu, The Second Affiliated Hospital of Nanjing Medical University, Nanjing, Jiangsu, China (e-mail: zhujg06@163.com).*

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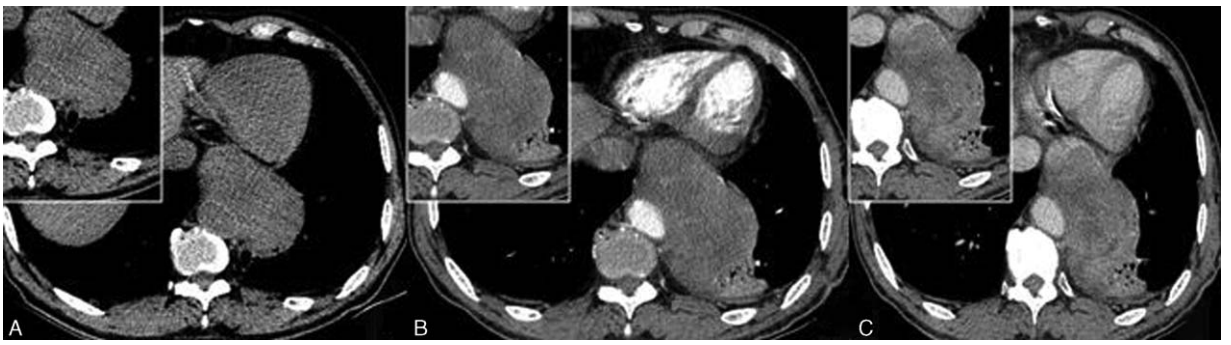

Figure 1. A, Unenhanced CT scan shows a mass with equal density in the posterior mediastinum. B and C, On contrast-enhanced CT images, the tumor presents moderate, heterogeneous enhancement. CT = computed tomography.

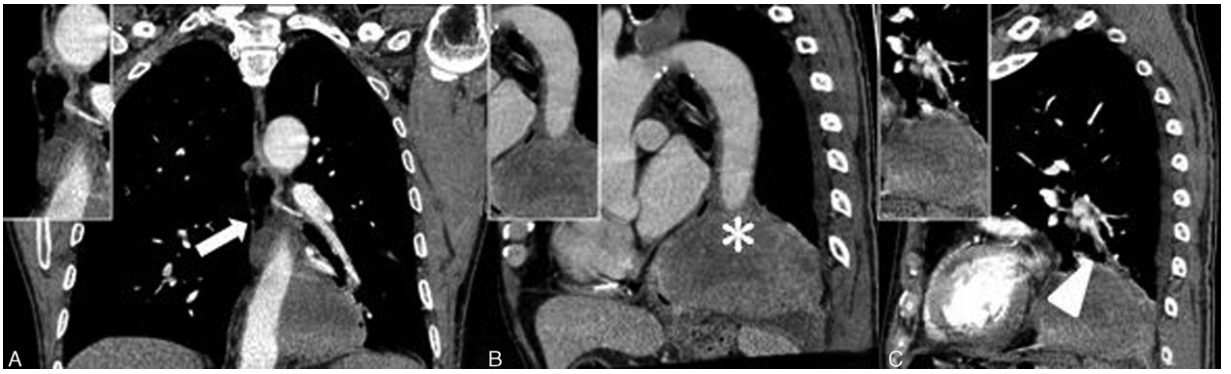

Figure 2. A, Contrast-enhanced CT images show the tumor encircles the esophagus (arrow). B, The descending aorta is compressed by the tumor (asterisk). C, Tracheal oppression by the tumor is presented on CT scan (triangle). CT = computed tomography.

percentage of Ki-67 antigen-positive nuclei, was less than 20%. The pathologic diagnosis was AMFB.

3. Discussion

Clinically, AMFB presents as a well-defined, painless mass occurring between the ages of 17 and 86 years (mean 45 years), with greatest diameters ranging from 3.8 to 25 cm.[4] In most female patients, AMFB arises in lower genital tract and is therefore easily noticed. Four cases of pelvic AMFB have been reported in Qiu et al's study.[4] These 4 slow-growing tumors reached a massive size and caused obstructive symptoms before they were detected. In the current report, the tumor of $6.9 \times 7.4 \times 9.3$ cm in size was located in the posterior mediastinum. It mechanically compressed the esophagus and caused dysphagia. To our knowledge, similar cases of AMFB originating from the mediastinum have not been reported in the previous published literatures.

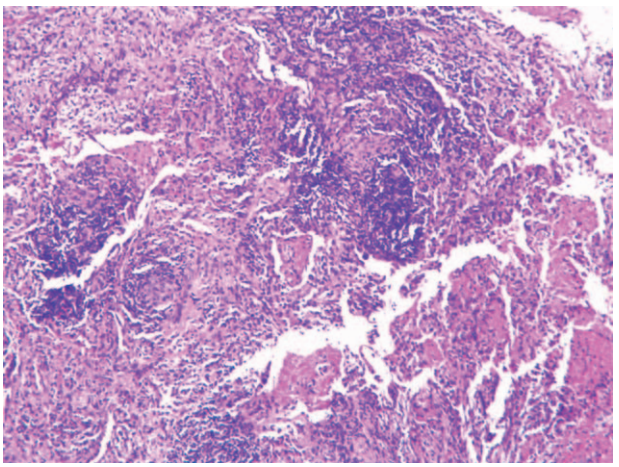

Figure 3. Spindle-shaped tumor cells cluster about blood vessels (H&E, ×100). H&E = hemotoxylin and eosin.

Histologically, AMFB is consisted of 2 basic components: small to medium-sized thin-walled vessels and round to spindle-shaped epithelioid cells, with prominent vascular component.[5] The tumor cells always exhibit a tendency for perivascular concentration, as presented in the current case. Immunologically, AMFB is almost uniformly positive for vimentin expression and frequently positive for expression of desmin. There is variable expression of muscle-specific actin and CD34, but it is typically negative for S-100 protein expression.[6] In the present case, tumor cells were positive for vimentin, desmin, and CD34, but there was no immunoreactivity for S-100 protein.

According to several published reports, the imaging features of AMFB have been summarized. Sonographically, 2 AMFB cases were manifested as solid cystic masses in Wang et al' study.[7] In contrast, the tumor appeared as a homogeneous, medium echogenicity mass with several vessels in Wolf et al's case report.[8] On CT imaging, AMFBs show as well-defined

homogeneous intermediate intensity and moderate to strong enhancement after contrast administration.[4] On magnetic resonance imaging (MRI), homogeneous hypointensity observed on T1 and T2-weighted images may be attributable to the fibrous structures.[8] In our current report, CT described a poorly circumscribed heterogeneous soft-tissue mass located in the posterior mediastinum. The tumor was moderately enhanced on contrast-enhanced CT, which may reflect the vascularity of the tumor.

Aggressive angiomyxoma (AAM), schwannoma, and solitary fibrous tumor (SFT) are considered in the differential diagnosis of AMFB. AAM is most likely to be confused with AMFB because both tumors share many common features. AAM is an aggressive neoplasm with infiltrative growth pattern. Small clusters of smooth muscle cells aggregating around blood vessels are a characteristic feature of AAM. The vessel walls of AAM are thick and large compared with AMFB.[6] After administration of intravenous contrast medium, AAM shows strong enhancement with swirled central strands of hypointense tissue caused by stretching of the fibrovascular stroma during protrusion.[9] Schwannoma is less vascular and more commonly expresses S-100 protein different from AMFB. Typical schwannoma is depicted as an inhomogeneous low-density mass with peripheral enhancement on CT scan.[10] AMFB is distinguished from SFT due to the spindle-shaped and bland appearance of the tumor cells, and also stronger positivity and more diffuse staining for CD34.[11] CT characteristics of SFT are nonspecific and include variable heterogeneous enhancement, calcifications, and areas of central necrosis. Hypointensity in T2-weighted images and spoke wheel pattern of enhancement are the typical imaging features of SFT.[12]

Surgical resection remains the primary treatment for AMFB, and radiation and chemotherapy play no role in the treatment.[13] Close postoperative monitoring and long-term follow-up management are recommended.[14]

In conclusion, our case adds to the experience with AMFB. This tumor represents a further example of unusual location different from conventional AMFB. The diagnosis of AMFB is based on several findings including clinical, radiological, and histological data.

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