

# Angiosarcoma originating in the anterior mediastinum

## A case report

Yan-Bin Tan, MD<sup>a,\*</sup>, Xin-Feng Yu, MD, PhD<sup>a</sup>, Jun-Qiang Fan, MD, PhD<sup>b</sup>, Jin-Fan Li, MD, PhD<sup>c</sup>

### Abstract

**Rationale:** Angiosarcomas are malignant vascular tumors, and angiosarcoma occurring in the anterior mediastinum is rare. Here we report a case of angiosarcoma that originated in the anterior mediastinum treated with surgery, followed by radiotherapy and synchronous chemotherapy.

**Patient concerns:** A 56-year-old female was admitted to our hospital with chest pain for 3 days. Chest computerized tomogram (CT) examination showed a heterogeneous mass in the anterior superior mediastinum, and after injection of contrast agent, the mass showed obvious heterogeneous enhancement. Magnetic resonance imaging (MRI) with T1 weighted image (T1WI) showed isointensity and T2 weighted image (T2WI) showed heterogeneous signal intensity, the mass showed an obvious heterogeneously enhancement after intravenous administration of contrast material.

**Diagnosis and interventions:** Surgical resection operation was carried out. According to its morphologic and immunohistochemic feature of tumor cells which expressing CD31, CD34, and ERG, the tumor was categorized as an angiosarcoma. After operation, the patient received radiotherapy and synchronous chemotherapy.

**Outcomes:** At present, 8 months postoperatively, no signs of recurrence have been observed.

**Lessons:** Although angiosarcoma in anterior mediastinum is rare, when a mass located in this area, a more careful immunohistological analysis should be performed to avoid overlooking the presence of angiosarcoma.

**Abbreviations:** CT = computerized tomogram, DWI = diffusion weighted image, HIV = human immunodeficiency virus, MRI = magnetic resonance imaging, T1WI = T1weighted image, T2WI = T2 weighted image.

**Keywords:** angiosarcoma, anterior mediastinum, radiotherapy, surgery, synchronous chemotherapy

## 1. Introduction

Angiosarcoma is an uncommon malignant tumor of endothelial cells. It can occur in anywhere in the body, but it is more frequently found in the skin, superficial connective tissue. Only a few cases have been found in deep soft tissue and the body cavity.<sup>[1]</sup> As we know, primary angiosarcomas arising in the thorax such as angiosarcoma of lung, heart and mediastinum are rare. And angiosarcomas that occur in the anterior mediastinum without an obvious vascular origin are extremely rare. Here we report a case of angiosarcoma that originated in the mediastinum

which was treated with surgery, followed by radiotherapy and synchronous chemotherapy. We obtained the informed consent for publication of the case from the patient and her husband.

## 2. Case report

A 56-year-old female was admitted to our hospital with chest pain for 3 days, accompanied with chest tightness and mild fever. The chest pain could be relieved after resting. She denied orthopnea, palpitations, night sweats, weight loss, and anorexia. She denied smoking, drinking, and the history of human immunodeficiency virus (HIV) and hepatitis B exposure, as well as occupational or recreational exposure to toxins.

The physical examination found no symptom or physical abnormalities. The laboratory examinations including blood routine test, coagulation function test, liver and kidney function test, urine routine test, and tumor markers were normal. Chest computerized tomogram (CT) showed that the lesion was located in the anterior superior mediastinum, with unclear boundary with superior vena cava and the right atrium. The size of lesion was 59 mm × 38 mm. After injecting of contrast agent, the mass showed a heterogeneous enhancement with the vast areas of necrosis (Fig. 1). Magnetic resonance imaging (MRI) showed the mass had isointensity on T1weight image (T1WI), heterogeneous signal intensity on T2 weight image (T2WI), and diffusion restriction on diffusion weighted image (DWI), the mass showed an obvious heterogeneously enhancement after intravenous administration of contrast material (Fig. 2).

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<sup>a</sup> Department of Radiology, <sup>b</sup> Department of Thoracic Surgery, <sup>c</sup> Department of Pathology, Zhejiang University School of Medicine Second Affiliated Hospital, Hangzhou, China.

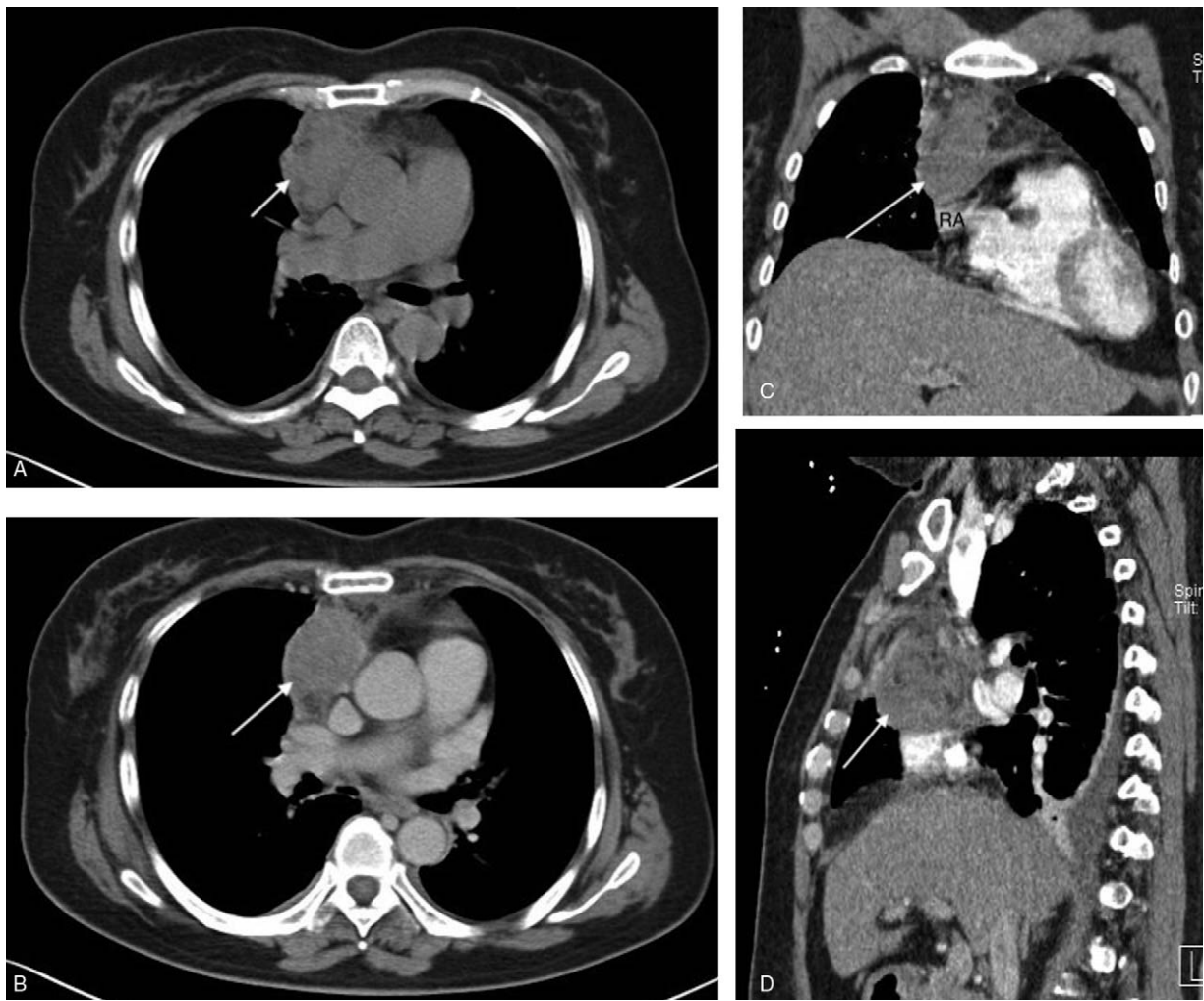
\* Correspondence: Yan-Bin Tan, Department of Radiology, Zhejiang University School of Medicine Second Affiliated Hospital, Hangzhou, China (e-mail: 2510009@zju.edu.cn).

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**Figure 1.** The CT scan of the mass (arrow) in the anterior mediastinum. (A) It is hypointensity precontrast enhancement, and the boundary is unclear. (B) The mass is obvious irregular enhanced after contrast enhancement. (C and D) Sagittal and coronal position of chest CT show the mass is abutting the superior vena cava (S) and the right atrium (RA). CT = computerized tomogram.

As there was no evidence of local or distant spread, surgical resection operation was carried. During the operation, a large tumor was seen, invading the pericardium and adherent to superior vena cava. Intraoperative pathological examination showed the spindle cell tumor with massive hemorrhage and necrosis. Immunohistochemical investigation revealed that the spindle cells expressing CD31, CD34, and ERG. According to its morphologic and immunohistochemical features, the tumor was finally diagnosed as an angiosarcoma (Fig. 3).

The patient was given a course of radiotherapy to the tumor bed and 4 courses of synchronous chemotherapy (cisplatin 50 mg qw) after surgery. At present, 8 months postoperatively, no signs of recurrence have been observed.

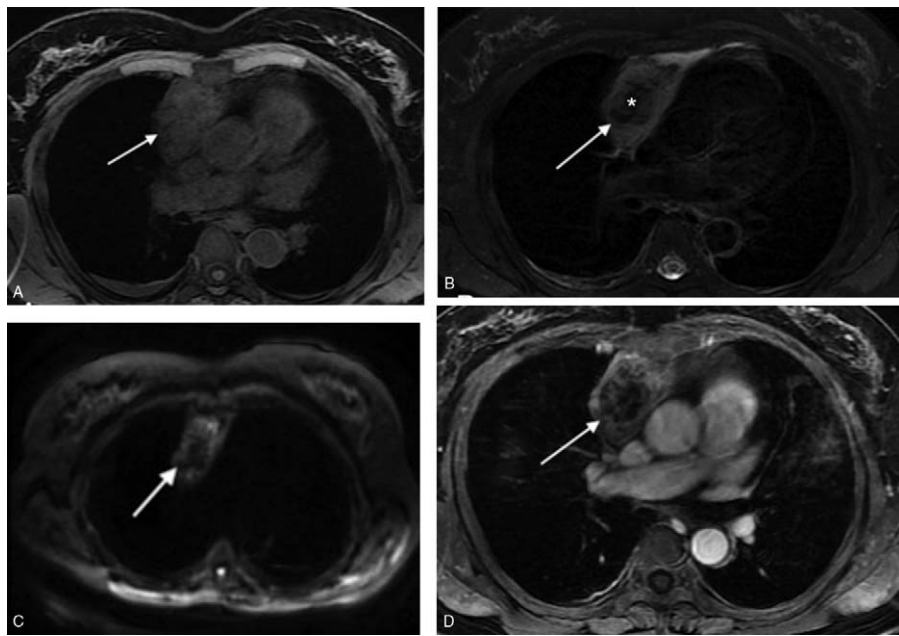
### 3. Discussion

Angiosarcoma is an uncommon sarcoma arising from the endothelium. A review of 300 cases of angiosarcomas reveals that one-third of angiosarcoma occurs in the cutaneous layers (primarily in the head and neck), one-fourth in the soft tissue, and the rest appears in various organs such as liver, spleen, breast,

bone, and heart.<sup>[1]</sup> In addition to the sporadic forms, there is a strong association of angiosarcomas occurring with radiation, environmental carcinogens, chronic lymphedema, or foreign bodies.<sup>[2]</sup>

Angiosarcomas typically occur in middle-aged adults. The clinical symptoms depend on their location. Mediastinal angiosarcomas manifest as a result of local mass effect, and chest pain is the usual presenting symptom. The symptom of the case we reported was chest pain. Angiosarcomas arising in the lung have been reported to cause diffuse alveolar hemorrhage.<sup>[3]</sup> And recently, Datta et al<sup>[4]</sup> reported a case of mediastinal angiosarcoma presenting as diffuse alveolar hemorrhage. They explained the likely cause of alveolar hemorrhage was alveolar metastases with probable back-pressure effect of mediastinal tumor causing obstruction to great vessels.<sup>[4]</sup>

Angiosarcomas of mediastinum has been rarely reported.<sup>[5,6]</sup> They mostly arise from the right atrium, pulmonary artery, or lung parenchyma. In a review of 1046 patients of mediastinal tumors over a 40-year period, found only 7 cases of angiosarcoma were reported.<sup>[6]</sup> However, both CT and MRI information are scarce in previous reviews. In our case, both CT

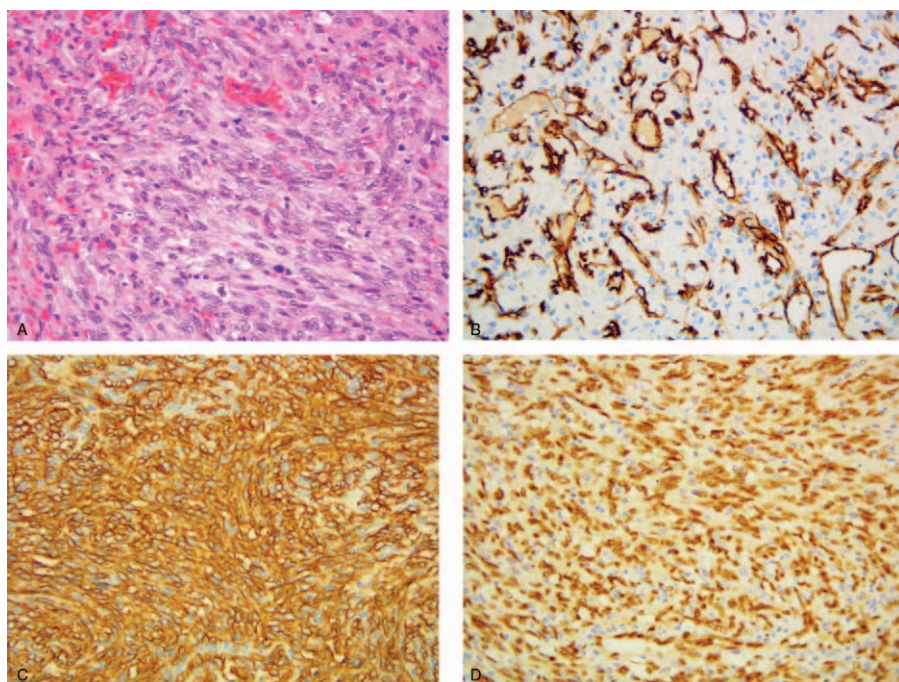


**Figure 2.** The MRI scan of the mass (arrow) in the anterior mediastinum. (A, B) Axial T1-weighted image (A) shows the mass is isointensity signal, and axial T2-weighted image (B) shows the mass is heterogeneous with area of low signal intensity which representing the hemorrhage (\*). (C) DWI shows the mass is diffusion restriction. (D) Axial gadolinium-enhanced T1-weighted image shows the mass is heterogeneous enhanced. DWI=diffusion weighted image, MRI=magnetic resonance imaging.

and MRI showed the tumor had an aggressive and infiltrative appearance. On CT, it had low attenuation with heterogenous enhancement postcontrast. On MRI, it is heterogenous due to the presence of intralesional hemorrhage and necrosis. Although

these imaging features suggest a malignant tumor, it is misdiagnosed as invasive thymoma before surgery.

Histopathological evaluation of an angiosarcoma is difficult, as it can exhibit variably atypical features ranging from mild



**Figure 3.** Microscopic appearance of the tumor. Hematoxylin-eosin stain ( $\times 400$ ) shows the immature vascular structures are formed with malignant epithelioid cells (A). Immunohistochemical staining ( $\times 400$ ) for CD31 (B), CD34 (C), and ERG (D) are positive.



nuclear hyperchromasia to highly pleomorphic cells with atypical mitotic figures.<sup>[7]</sup> Immunohistologically, staining for vascular marker factor VIII-related antigen, CD31 and CD34, are positive in angiosarcomas. In the case we reported, immunohistological staining of CD3, CD34, and ERG were positive. So, there was enough evidence from the various tests to diagnose it as an angiosarcoma.

The prognosis of angiosarcoma is poor, and 5-year survival rates is about 24%, respectively.<sup>[8]</sup> For some unresectable cases, the median survival time is only 7.3 months.<sup>[9]</sup> Because of the rarity of this disease, there are no accepted guidelines. Surgical resection with or without adjuvant irradiation or chemotherapy has been the main treatment modality for this disease.<sup>[9,10]</sup> It has been reported that the aggressive surgical resection and additional radiation therapy for close margins can result in long-term survival.<sup>[9]</sup> With surgery and chemotherapy, the survival was reported in one series to be 17% .<sup>[8]</sup> Comparing these survival times to those of angiosarcomas in other locations such as the deep soft tissue (median survival, 20 months) or the heart (median survival, 9–12 months), angiosarcomas of the anterior mediastinum seem to pursue a less aggressive clinical course.<sup>[7]</sup> Until now, the patient reported in this study shows no signs of recurrence for 8 months. Hence the treatment in our patient had a beneficial outcome.

#### 4. Conclusion

Angiosarcoma in anterior mediastinum is rare, but it should be included in the differential diagnosis of anterior mediastinum neoplasms. Both findings of CT and MRI may be nonspecific.

Immunohistological staining with vascular markers expression are important for accurate diagnosis.

#### Author contributions

**Data curation:** Yan-Bin Tan, Jun-Qiang Fan, Jin-Fan Li.

**Writing – original draft:** Yan-Bin Tan.

**Writing – review & editing:** Xin-Feng Yu.

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