



A Case of Primary Pulmonary Angiosarcoma with Pleural Metastasis

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Abstract

Angiosarcomas are a subtype of soft tissue sarcomas which are aggressive, malignant endothelial-cell tumors of vascular or lymphatic origin. Even though the lung is a vascular organ; it is usually not the primary site for vascular neoplasms. The lungs are more often the site of metastasis from extrapulmonary tumors. We report a 26 year-old previously healthy gentleman who presented with chest pain. His chest images showed multiple bilateral nodules and nodularity of the left pleura. He underwent Video Assisted Thoracoscopy (VATS) and nodule biopsy which showed Angiosarcoma.

Introduction

Angiosarcomas are a subtype of soft tissue sarcomas which are aggressive, malignant endothelial-cell tumors of vascular or lymphatic origin [1]. It can arise from any part of the body with skin and soft tissue being the most common sites involved [2]. It is a rare malignant tumor which accounts for less than 1% of all sarcomas and generally 2% to 3% of all soft tissue tumors are vascular sarcomas [2-7]. Most Angiosarcomas arise spontaneously, but there are a few reports of malignant transformation within pre-existing benign vascular lesions [1,3]. It is included in the latest WHO classification of vascular sarcomas under the highly aggressive neoplasms [4,5]. Even though the lung is a vascular organ, it is usually not the primary site for vascular neoplasms. The lungs are more often the site of metastasis from extrapulmonary tumors [8]. Because of the rarity of these lesions, the clinical and radiologic features are often mistaken for other, more common processes until the vascular nature of the disease is confirmed by histologic examination [8]. Pulmonary angiosarcoma has a high rate of recurrence and is frequently disseminated at the time of diagnosis [2,9].

In our review, we were able to find at least 60 cases of primary pulmonary angiosarcoma reported in the English literature. Most likely there are more cases that were not reported. We report a 26 year-old previously healthy gentleman who was found to have primary pulmonary angiosarcoma with pleural metastasis.

Case Presentation

A previously healthy 26 year-old gentleman presented with 3 months history of left sided pleuritic chest pain. He gave history of dry cough and mild shortness of breath with exertion. During his illness he developed fever and weight loss of around 6 Kg. He is a lifelong non-smoke. He works as a pharmacist and has no significant exposures. His Family history was positive for different kinds of malignancies. Review of systems was unremarkable. On examination he was tachypneic but his oxygen saturation was 99% on room air. He was febrile with a temperature of 39°C and tachycardic. His chest examination showed an area of dullness at the left lower zone with decrease breath sounds at the same area with bronchial breathing. He had no palpable lymph nodes. The rest of his examination was unremarkable.

He was investigated in a different hospital (prior to being referred to our center) and was found to have a left pleural effusion and nodules on the right side. He had left sided thoracentesis more than once which was repeatedly hemorrhagic. Cytology from the effusion was inconclusive. PPD testing was negative. In our hospital, his chest X-ray showed left lower lobe opacity with obliteration of the left CP angle and nodules on the right lung. A chest ultrasound showed hyperechogenicity in the left pleural space but no fluid to be tapped. His chest CT showed multiple bilateral nodules and nodularity of the left pleura with dense material in the pleura suggestive of a hematoma. CT also showed lymphadenopathy in the hilum and mediastinum. He underwent Video Assisted Thoracoscopy (VATS) and nodule biopsy.

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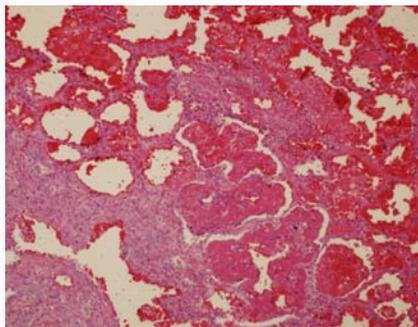


Figure 1: Lower power view showing dilated channels filled with blood.

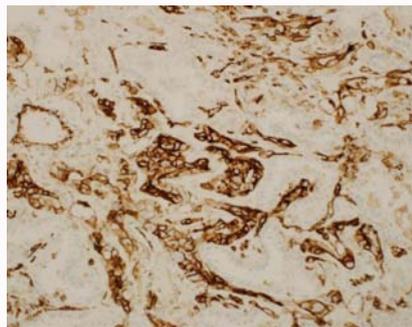


Figure 3: Epithelioid cells expressing CD31.

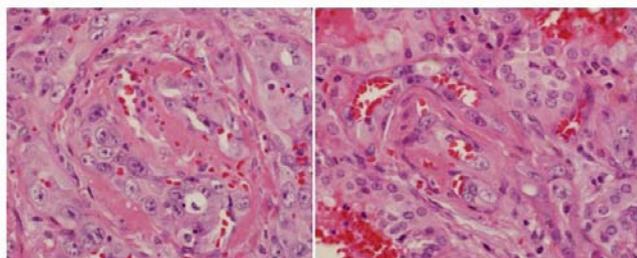


Figure 2: a) The lesion consists of clusters of high grade rounded epithelioid endothelial cells with prominent nucleoli, b) some of which contain intracytoplasmic lumen with red blood cells.

The specimen was reviewed by three of our pathologists and the final diagnosis was Angiosarcoma. A CT of the abdomen and pelvis showed no evidence of peripheral metastasis and a bone scan showed no evidence of bone metastasis. He underwent VATS and evacuation of the hematoma as it was thought that it was the source of his sepsis. Despite the evacuation of the hematoma, the fever did not subside. It was then assumed that his fever was mostly secondary to his underlying illness, as his cultures were repeatedly negative. After recovery he was transferred to The Medical Oncology team for palliative chemotherapy.

Discussion

Angiosarcomas represent less than 1% of all sarcomas and develop most often in the skin, soft tissue, or liver. They may be associated with previous radiation treatment; environmental carcinogens like vinyl chloride, thorotrast, or phenylethyl hydrazine, foreign body material or lymphedema. The lungs are more often the site of metastasis from extrapulmonary tumors, most frequently from the heart and the pulmonary artery trunk. In the lung, primary angiosarcomas are extremely uncommon [8,10-17].

The age of patients affected ranges between 22 to 79 years (mean, 54 years) with a male to female ratio of 3:1 [8]. Primary pulmonary angiosarcomas and metastatic angiosarcomas in the lung have similar symptomatology and radiologic features [2,18]. The most common presentation clinically includes hemoptysis, shortness of breath and weight loss. Chest pain has also been reported as a presenting symptom as more than 14 of the cases we found reported in the literature presented with chest pain similar to the patient we are reporting [9,18-28]. There have been reported cases of pulmonary hemorrhage as a presenting symptom. Radiologically, pulmonary angiosarcomas usually present with lung nodules on chest CT scan. The CT scan of pulmonary angiosarcoma has a distinctive appearance

of soft tissue attenuation surrounded by ground glass attenuation which is also known as the halo sign [23,29]. The halo could be due to hemorrhage in the surrounding lung from the nodule [23,29]. There have been reports of primary pulmonary angiosarcoma being detected by PET-CT scan [30-32]. Primary pulmonary angiosarcoma has a very poor prognosis with the longest survival period reported to be 39 months [9,26]. Treatment with radiotherapy and recombinant interleukin-2, chemotherapy using ifosfamide-doxorubicin combination, pneumonectomy, lobectomy and radiotherapy have been used as treatment options with variable results but the outcome remains poor [9,22,26].

Pleural metastasis from a primary pulmonary angiosarcoma has been reported. Of the cases we reviewed (60 cases); at least 12 were reported to have pleural involvement. It seems the yield from pleural fluid cytology is low as we could not find any report diagnosing a patient based only on the pleural fluid. Ayabe et al. [9] attempted pneumonectomy of primary pulmonary angiosarcoma with malignant effusion and intrapleural hypotonic hyperthermic chemotherapy and the patient was alive at the time of their report (21 months). Primary pleural vascular tumors (including angiosarcoma) are even rarer [33]. Even though we could not document it, we do not suspect that this patient had a primary metastatic pleural vascular tumor.

Conclusion

Primary Pulmonary angiosarcoma is a rare disease with a very poor prognosis and no definite treatment so far. More cases have been reported in recent years with different therapeutic modalities, none of which proving to be effective. Pleural involvement is not an uncommon feature in such patients.

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