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Chest

Pulmonary epithelioid angiosarcoma responsive to chemotherapy: A case report

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ABSTRACT

Primary pulmonary epithelioid angiosarcoma (AS) is an extremely rare cancer with a poor prognosis. The presenting symptoms and imaging results are nonspecific and hold similarities with more common lung pathology, contributing to missed or delayed diagnosis. Complementing radiological imaging with patient information, such as presenting symptoms and exposures, is important for early consideration of pulmonary epithelioid AS. Even with supportive imaging findings and clinical suspicion for pulmonary epithelioid AS, the most reliable and definitive method for diagnosis is through immunohistochemistry. We describe the case of a 65-year-old patient who presented with dyspnea, cough, and hemoptysis in whom pauci-immune vasculitis was initially suspected before immunohistochemical diagnosis of primary pulmonary epithelioid AS. Due to the rarity of this disease, treatment options have not been well-studied and consist of any combination of surgical resection, chemotherapy, and radiation therapy. Although typically poorly responsive to chemotherapy, our patient achieved a reduction in size of his pulmonary nodules after a course of steroids followed by cyclophosphamide and was later maintained with gemcitabine and docetaxel until his death nearly a year after presentation.

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Introduction

We report the case of a 65-year-old male with primary pulmonary epithelioid angiosarcoma (AS), a lethal neoplasm of extreme rarity that originates from endothelial cells of small vessels and accounts for approximately 0.001% of all cancers. Primary pulmonary epithelioid AS is seen in adults and has a marked predominance in males, occurring 6 to 9 times more often in men than women by some study estimates [1,2]. Presenting signs and symptoms commonly include chest pain, dyspnea, hemoptysis, cough, and weight loss. Among the differential diagnoses for primary pulmonary epithelioid AS are more common pulmonary diseases, such as metastatic disease

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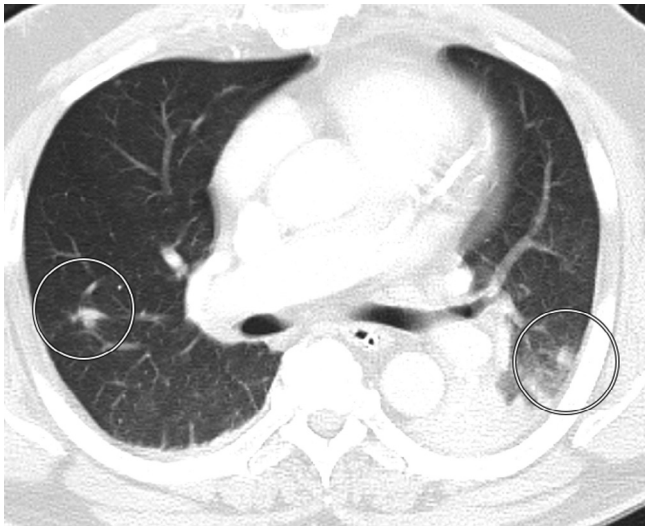

Fig. 1 – Representative computed tomography image in lung windows showing small bilateral pulmonary nodules. There is adjacent consolidation in the left lower lobe.

and mesothelioma. Imaging and histology of primary pulmonary epithelioid AS are often suggestive but nonspecific, so a strong clinical suspicion followed by immunohistochemical confirmation of the endothelial tumor origin is necessary. This neoplasm is aggressive and often detected late in the disease course, thus associated with a poor prognosis.

Case

A 65-year-old male with a past medical history significant for type 2 diabetes mellitus, coronary artery disease status post two-vessel bypass, hypertension, and hyperlipidemia presented to his primary care physician complaining of shortness of breath and cough for which he was prescribed an oral steroid and albuterol. The patient denied a history of smoking or asbestos exposure, intravenous drug use, or human immunodeficiency virus exposure. The patient had worked in a coal mine for approximately 30 years, but past surgical, family, and social histories were otherwise noncontributory. Two days later, he presented to an outside hospital with diffuse alveolar hemorrhage and was electively intubated for bronchoscopy. His initial hemoglobin was 9.4 g/dL. Urinalysis revealed no blood or protein with a creatinine of 0.77 mg/dL. Liver function tests were not performed. Computed tomography (CT) of the chest with contrast was performed as part of the initial workup at an outside hospital and demonstrated bilateral pulmonary nodules (Fig. 1). The largest nodule measured 12-13 mm in the costophrenic sulcus and was concerning for metastatic disease. Bibasilar atelectasis was present and more pronounced in the left than right lower lobe. No other abnormalities were noted. He was extubated but later reintubated for worsening hemoptysis. At this point, he received blood transfusions and underwent embolization of his left lower lobe by interventional radiology.

Approximately 1 week later, he experienced extensive bleeding from his endotracheal tube that slowed but persisted after embolization. A repeat CT of the chest without contrast revealed worsening bilateral lower lobe atelectasis, left lower lobe mucous plugging with an enlarging focus of cavitation, and bilateral perihilar ground-glass opacities, one of which was new and cavitary. In addition to metastasis, septic emboli and vasculitis were also in the differential. Pulse methylprednisolone was started at 500 mg intravenously every 12 hours for 3 days, and he was extubated approximately 1 week later. A subsequent CT of the chest without contrast demonstrated that the lower lobe cavitary component had resolved, albeit with consolidation concerning for pneumonia and areas of hemorrhage. The bilateral pulmonary nodules were stable, and the previously new cavitary nodule was not evident. These findings indicated a response to treatment and were considered suggestive of granulomatosis with polyangiitis. During hospitalization in the intensive care unit, blood cultures remained negative. Although initial bronchoalveolar lavage (BAL) specimens were negative, subsequent BAL cultures revealed *Enterobacter* species for which the patient received cefepime. Serology remained negative for an infectious source, and results from antigen testing for *Cryptococcus*, *Histoplasma*, *Aspergillus*, and *Coccidioides* were negative as well. The (1→3)-β-D-Glucan assay was initially inconclusive but negative on repeat testing. He was believed to have antineutrophil cytoplasmic antibody-negative granulomatosis with polyangiitis given his presentation and aforementioned suggestive findings, and cyclophosphamide at a dose of 150 mg by mouth daily was initiated. He was transferred from the intensive care unit and discharged to home several days later in stable condition with 60 mg oral prednisone.

He attended follow-up appointments with rheumatology and pulmonology and continued cyclophosphamide therapy. The patient was recovering well and experienced only a single episode of hemoptysis. He did complain of intermittent dyspnea upon exertion and was given an albuterol metered-dose inhaler. An outpatient follow-up CT scan of the chest without contrast, approximately 2 weeks after discharge, revealed stable bilateral pulmonary nodules. The most suspicious nodule was in the right lower lobe and measured 2.2 cm with spiculated margins and an irregular shape. A 2.2-cm left lower lobe nodule of fluid density was new and suggestive of a bronchocele, while the ground-glass infiltrates had resolved. By 3 months after discharge, his episodes of blood-tinged saliva progressed to worsening hemoptysis. An outpatient follow-up chest CT without contrast at this time showed multiple persistent nodules with increasing size of the nodules in the upper lobes and decreased size of most of the nodules in the lower lobes. No adenopathy or effusion was evident. A third outpatient CT of the chest without contrast was obtained, revealing relatively stable bilateral pulmonary nodules with an increase in size of the dominant right lower lobe pulmonary mass to 3.4 × 2.7 cm (Fig. 2). The surrounding ground-glass halo was suggestive of hemorrhage.

Bronchoscopy with transbronchial fine needle aspiration from the right upper lobe and core biopsy demonstrated malignant cells suggestive of sarcomatoid carcinoma or epithelioid angiosarcoma. Immunohistochemistry of the biopsy specimen was positive for AE1/3, pan-cytokeratin, CK7, and epithelial

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