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From Head to Hilum: Angiosarcoma with Pulmonary Metastasis

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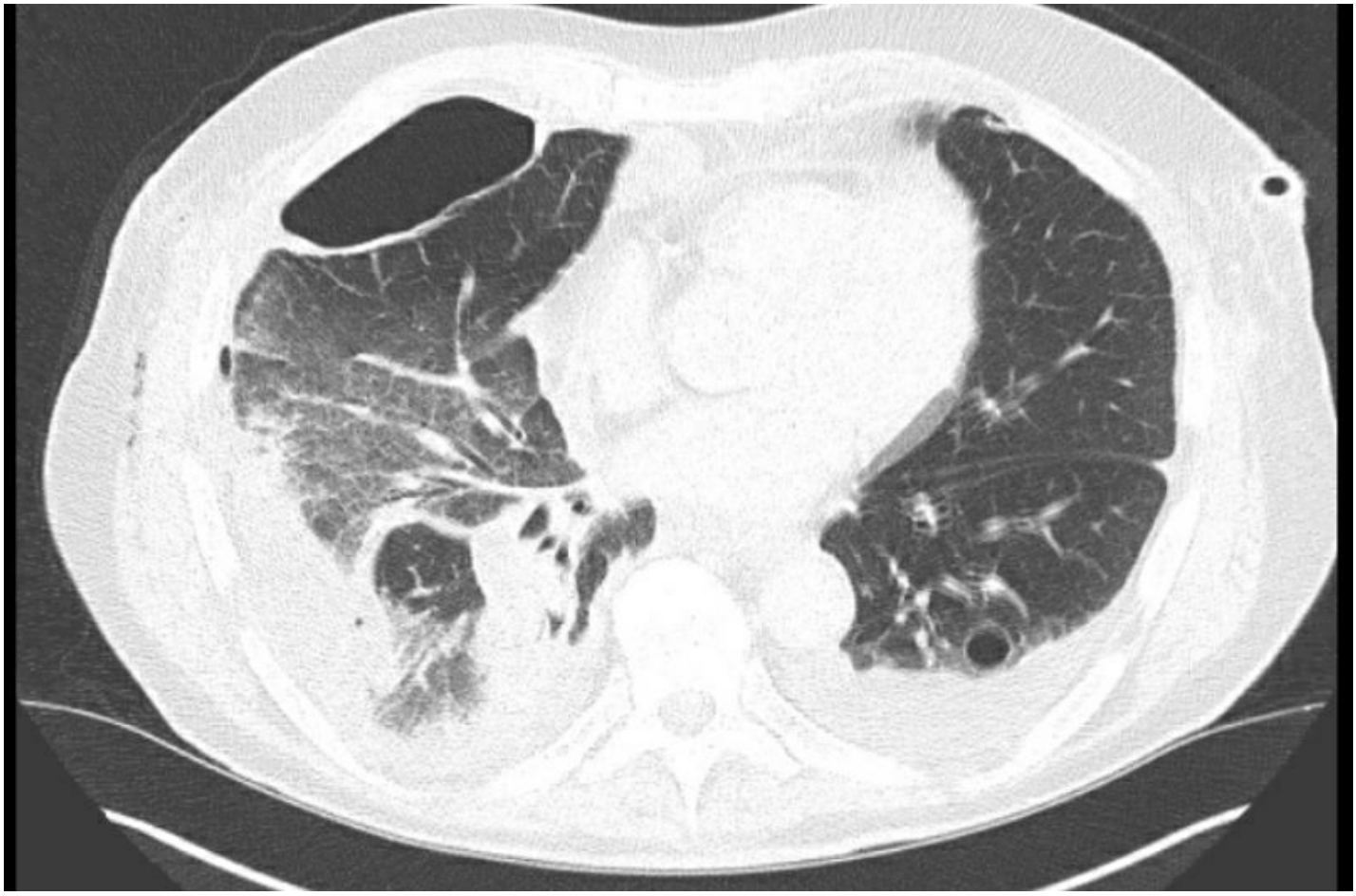
With an estimated five-year survival of less than 40 percent, angiosarcomas are rare and often misdiagnosed fatal tumors originating from blood or lymphatic vessels with a predilection for the head and neck. They account for 1 percent of all soft tissue sarcomas and are often mistaken for benign mimickers leading to the late-stage diagnosis when the body is already riddled with metastatic disease with the lung being the most common site of metastases. This case illustrates the myriad of pulmonary complications associated with this uncommon but fatal vasoformative malignancy. The patient is a 73-year-old male diagnosed with angiosarcoma of the right scalp status post excision, reconstruction, and adjuvant proton radiotherapy. He presented to the hospital 8 weeks after treatment completion with new onset acute hypoxic respiratory failure secondary to bilateral pleural effusions as well as bilateral hydropneumothoraces. He underwent bilateral thoracenteses as well as right and left chest tubes with a Heimlich valve. On CT chest, he was found to have resolution of the effusions and pneumothoraces but an increase in the number of pulmonary cysts, nodules and ground glass opacities. Cytology of the effusions was positive for metastatic angiosarcoma. When angiosarcoma is diagnosed, it is imperative that clinicians familiarize themselves with the pulmonary manifestations of metastasis as they indicate rapid progression of the disease as well as worse prognosis. Patients can present with hemoptysis, cough, pleuritic chest pain or dyspnea resulting from several apathologies ranging from diffuse alveolar hemorrhage to pneumothorax to pleural effusion. Imaging is often non-specific and can show a pneumothorax (often recurrent due to cyst rupture), effusions, irregular pulmonary cysts or nodules as well as reticulonodular opacities. Due to the multitude of symptoms associated with metastasis, patients often get misdiagnosed with vasculitis, COPD flares or infections such as non-mycobacterium TB, further postponing appropriate chemotherapies. Angiosarcoma in it of itself is an often missed or delayed diagnosis. When the disease process is confirmed, one must be aware of the high rate of metastasis with the lung being the most likely site. Due to the high mortality rate, it is imperative that pulmonary symptoms not be confounded with the aforementioned less lethal etiologies. Early detection and treatment with excision, radiation therapy and chemo will allow for higher rates of disease-free survival.



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