Fatal Hemoptysis: The Sum of all Fears

Smit Mehta DO[1], Colin Eash DO[1], Fadi Aldaas MD[2]

[1]Internal Medicine, University of Illinois - Advocate Christ Medical Center, Oak Lawn, Illinois, United States
[2]Pulmonology & Critical Care, University of Illinois - Advocate Christ Medical Center, Oak Lawn, Illinois, United States

BACKGROUND

Angiosarcoma is a rare malignant neoplasm of the vascular endothelium that accounts for approximately 2% of all soft tissue sarcomas. Primary lung and cardiac angiosarcoma are even rarer entities with incidence rates at 0.003% and 0.0001%. The majority of literature describes a dismal prognosis w/ median survival of 6 months. Standard chemotherapy, radiation, and surgery have been consistently unsuccessful. Today we present a case of fatal hemoptysis from primary cardiac angiosarcoma with metastasis to the lungs.

CASE PRESENTATION

59 year-old man w/ past medical history of hypertension, heart failure with preserved ejection fraction, atrial flutter on apixaban who presented to the hospital with a chief complaint of hemoptysis. Several months prior to admission patient had underwent an radiofrequency ablation for atrial flutter that was riddled with complications including hemopericardium and pericardial tamponade requiring a pericardial window. Upon readmission, blood-tinged sputum had been present for several weeks prior to frank hemoptysis. CT pulmonary angiography revealed extensive ground glass opacities in both lungs (Exhibit A), a large pericardial effusion (Exhibit B/C) and a 4.5cm lobular mass in the epicardial fat tissue along the right heart border (Exhibit C).

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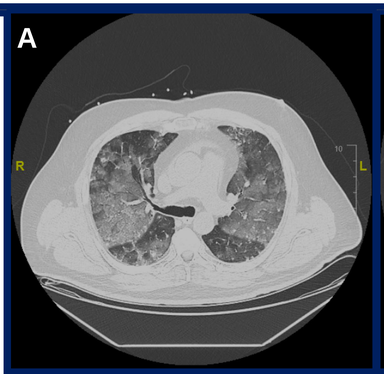

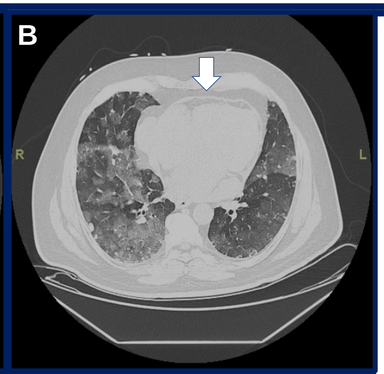

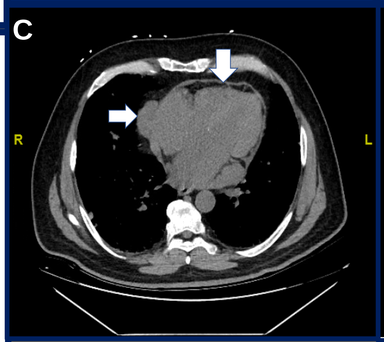

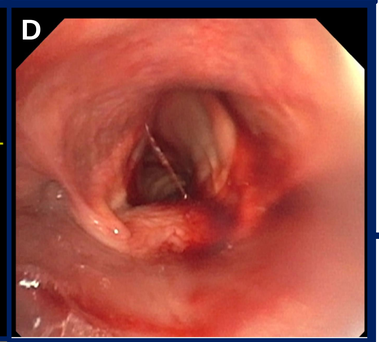

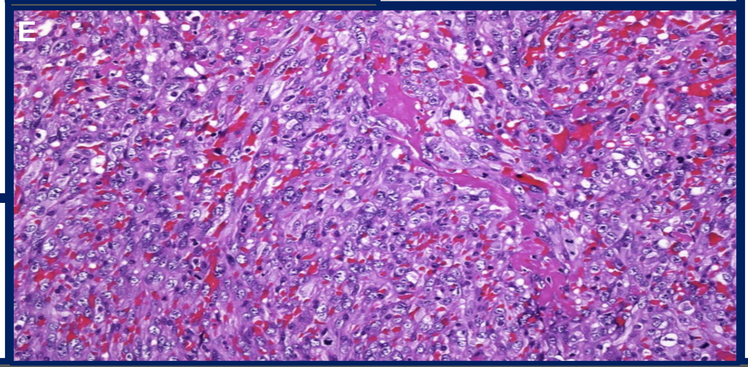

HOSPITAL COURSE

He underwent bronchoscopy which confirmed diffuse alveolar hemorrhage by serial aliquots (Exhibit D), followed by a video-assisted thoracoscopic surgery and was transferred to the intensive care unit. Massive hemoptysis ensued requiring emergent coil embolization with minimal improvement. With vasculitis on the differential, empiric plasmapheresis was started with pulse dose steroids with no response. Worsening pulmonary infiltrates with progression to acute respiratory distress syndrome and multi-organ failure quickly followed. Pathology results from thoracoscopic surgery returned during final days of hospital course showing malignant invasive cardiac angiosarcoma with metastatic involvement of bilateral lung parenchyma (Exhibit E). Patient eventually succumbed to his illness and expired with hospice care.

CONCLUSION

Primary cardiac angiosarcoma with metastatic lung involvement is an extremely rare disease with a poor survival time from diagnosis. The median survival of patients is an astonishing 1.5 months. Median overall survival of a patient with primary lung angiosarcoma is not much better at 5 months. We highlight the importance of this case as a rare clinical entity that requires a multi-disciplinary approach with early recognition and intervention in hopes to provide effective treatment. To date however these tumors have a very aggressive clinical course and a grim prognosis despite maximal medical therapy.