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Hemoptysis as the first sign of angiosarcoma: an extremely aggressive cardiac tumor

Short title: Hemoptysis as the first sign of cardiac angiosarcoma

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A 19-year-old male was admitted to the hospital with a two-month history of hemoptysis. Previously healthy, with no significant family history, he was initially examined for lung pathology. On admission, a chest x-ray revealed disseminated densifications in the lungs. Computer tomography (CT) showed subpleural opacifications and multiple, bilateral solid nodules with ground glass opacities. (FIGURE 1A-B) The nodules were more dispersed in the peripheral lobules, which is suggestive of neoplastic spread [1]. The chest scans also showed an irregular mass in the right atrium. The patient was transferred to the cardiac surgery department. Transthoracic echocardiography (TTE) revealed a 70x45 mm heterogeneous lobulated tumor in the right atrium infiltrating the free wall of the right atrium, right ventricle and tricuspid valve, suggestive of malignancy. Cardiac magnetic resonance demonstrated inhomogeneous late gadolinium enhancement of the mass infiltrating the pericardium (Figure 1C-D). Laboratory tests showed elevated d-dimers, liver enzymes and fibrinogen, neoplastic biomarkers were within normal ranges. The most probable diagnosis was a primary cardiac neoplasm with metastatic spread; pulmonary lesions were highly suggestive of metastases of angiosarcoma. The possibility of operation was precluded due to the anatomical location and infiltrative character of the tumor. Biopsy of the lesion was necessary to implement any systemic treatment. Nevertheless, the patient refused this procedure and asked for discharge.

The patient was readmitted to the cardiology department two weeks later with severe dyspnoea, weakness and excessive hemoptysis. TTE confirmed enlargement of the lesion to 80x70 mm (FIGURE 1E-F). Due to hemoptysis the patient required numerous blood transfusions and intensified pain therapy. Biopsy of the tumor was eventually performed and poorly differentiated angiosarcoma cells classified as G3 were found. The patient was immediately transferred to the oncological center to undergo palliative chemotherapy. During transport his condition got worse and he was admitted directly to the Intensive Care Unit with
a suspicion of pulmonary embolism. After 2 days the patient died due to multiple organ
dysfunction and septic shock.

Cardiac angiosarcomas are a rare pathology with an average survival period of 7 months if
diagnosed in the metastatic stage [2]. Lesions of the right heart have a poorer prognosis
compared to those on the left. They are described as infiltrative, bulky, heterogeneous
lobulated masses with areas of hemorrhage and necrosis on echocardiography. [3] Surgical
resection is essential, and both complete and partial resection contribute to longer survival.
Adjuvant chemotherapy is used due to frequent metastases and its efficacy of reducing tumor
tissue. Combining surgical resection, chemotherapy and radiotherapy can extend survival time
up to 3 years. [4]

If the tumor is not resectable, biopsy and histopathological analysis is crucial to implement
systemic treatment. This case exemplifies that if angiosarcoma is suspected, the biopsy must
not be postponed. The course of disease is very aggressive with extensive progression in
tumor mass seen after two weeks. A prompt diagnostic workup is a vital factor for fast
implementation of targeted therapy and requires involvement of a multi-specialised team
[4,5].
References:


Figure 1. Multimodality imaging of cardiac angiosarcoma: A - X-ray of the lungs showing diffuse ground-glass opacities (black arrows) B - Computed tomography of the lungs showing ground glass opacities in the center and periphery (white arrows) and subpleural opacifications (black arrows); C - initial TTE, subcostal view showing a large right atrial tumor with a possible thrombus on the periphery; D - T1-weighted magnetic resonance imaging showing lesion in right atrium of the heart (thick arrow) and metastatic lesions in both lungs (thin arrows); E – TTE after 2 weeks, apical 4-chamber view showing the tumor mass enchroaching RA and infiltrating the free wall; F - TTE after 2 weeks, subcostal view, showing the enlargement of the tumor mass. Abbreviations: RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium; TTE – transthoracic echocardiogram