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CASE REPORT

Pulmonary artery sarcoma: a rare thoracic tumor frequently misdiagnosed at presentation

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Abstract

This case illustrates a rare but important differential diagnosis of pulmonary emboli in the field of thoracic oncology, that of pulmonary artery sarcoma. It describes particular clinical features that may raise suspicion of this tumor in cases of suspected pulmonary emboli, and highlights novel radiological modalities and tissue sampling techniques in such cases. Surgical resection, as part of multi-modality therapy, is the cornerstone of treatment that has seen survival dramatically improve in recent years for patients with this rare cancer.

Introduction

Acute pulmonary embolus is a disease commonly encountered by hospital physicians. There is, however, a rare but important differential diagnosis that must always be considered in such cases, that of pulmonary artery sarcoma. These two diseases have very similar presenting symptoms, albeit often with subtle differences that can identify those patients that require more detailed investigation. The following report illustrates such a case and describes novel methods of achieving a diagnosis.

Case history

A 61-year-old female patient presented with a two-month history of progressive breathlessness, weight loss, fatigue, and fever. Routine laboratory blood tests revealed a normocytic anemia, thrombocytosis, leucocytosis, and raised C-reactive protein (CRP). The d-dimer was significantly raised at 6335 ug/mL (reference range [0–500]). Computed tomography (CT) of the thorax demonstrated abnormal opacification within the right pulmonary artery (RPA) (Fig 1). The patient was initially diagnosed with pulmonary emboli and anticoagulation treatment commenced. Despite anticoagulation, both the symptoms and radiological abnormalities were unchanged one month later and she was referred to a tertiary centre for evaluation. Positron emission tomography (PET)-CT revealed abnormal [^{18}F]-2-fluoro-deoxy-D-glucose uptake within the RPA (maximum standardised uptake value [SUVmax] 8.6, Fig 2). A pulmonary angiogram, performed by the interventional radiology team, confirmed an obstructing RPA lesion. Catheter aspiration sampling provided tissue samples that demonstrated the presence of poorly differentiated large atypical cells, positive for vimentin (diffuse and strong) and CD31 (focal). Epithelial (AE1/3, MNF-116, Cam 5.2, cytokeratin [CK] 7, and CK 20), muscle differentiation (desmin and smooth muscle actin), and melanocytic markers (s-100) were negative. These features were in keeping with a high-grade pleomorphic sarcoma of the pulmonary artery. The patient underwent a surgical resection plus right pneumonectomy. Macroscopic examination of the resected tissue confirmed the presence of a

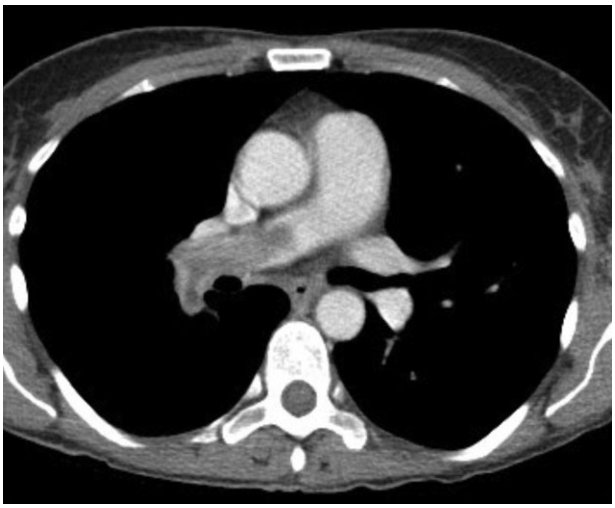

Figure 1 Computed tomography of the thorax demonstrating abnormal opacification within the right pulmonary artery.

smooth, friable tumor occluding the right pulmonary artery near its origin, extending into the lobar and segmental arteries in the right lung (Fig 3). The tumor appeared to arise from the endovascular surface, but the adventitial surface appeared smooth and intact. Microscopic examination confirmed a malignant tumor arising from the tunica intima. There were occasional foci of invasion into the tunica media but the tunica adventitia was uninvolved. Tumor emboli were present in the distal arterial branches. Immunohistochemistry revealed similar appearances to the previous sample confirming a diagnosis of intimal pulmonary artery sarcoma (Fig 4). Given the propensity for this tumor to metastasize and the presence of tumor emboli in the resected right lung, four cycles of adjuvant chemotherapy were recommended and subsequently completed. Three years on, the patient remains under follow-up with regular surveillance imaging. There are

Figure 3 Pneumonectomy specimen with transverse slices revealing tumor in the right pulmonary artery and its branches.

currently two left lung nodules and mediastinal soft tissue at the resection site being monitored closely, but to date there is no clear evidence of disease recurrence.

Discussion

Pulmonary artery sarcomas are extremely rare thoracic tumors, with approximately 300 cases reported to date.[1,2] Delay in diagnosis is frequent with pulmonary emboli the most common misdiagnosis.[1–5] Clinical features that may raise suspicions of pulmonary artery sarcoma in patients with suspected pulmonary emboli include constitutional symptoms, such as fever and weight loss, clubbing, and elevated inflammatory markers, such as erythrocyte sedimentation rate, and CRP. For patients in whom such suspicions have been raised, magnetic resonance imaging with gadolinium contrast can effectively differentiate between tumor and

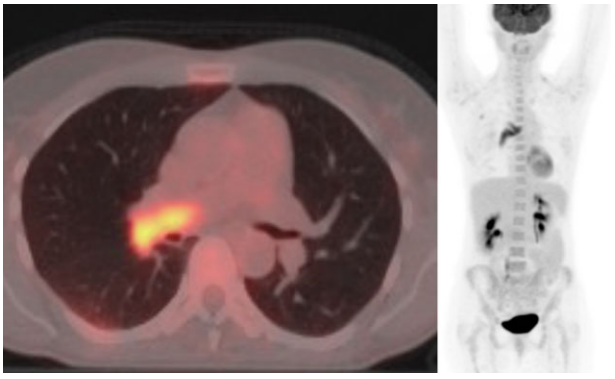

Figure 2 Positron emission tomography-computed tomography demonstrating abnormal fluorodeoxyglucose uptake within the right pulmonary artery.

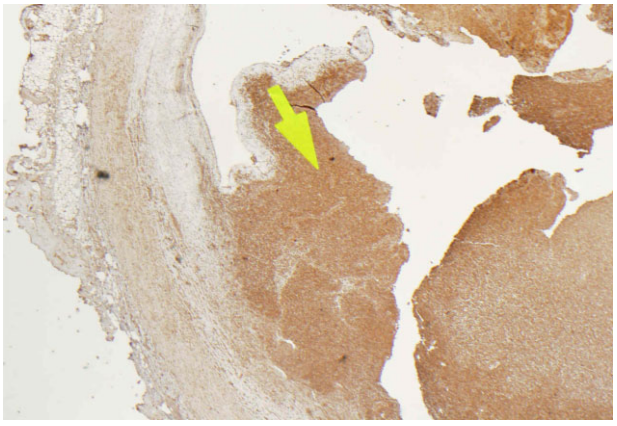

Figure 4 Pulmonary artery cross-section with endovascular tumor filling the vascular lumen and tumor cells staining positive for vimentin (10 × magnification).

thrombus.[6] In addition, as this case illustrates, PET-CT can perform a similar function and is a useful imaging modality. A previous study by Ito *et al*. demonstrated a significant difference in the mean SUVmax between three patients with pulmonary artery sarcoma and ten patients with pulmonary emboli (7.63 ± 2.21 vs. 2.31 ± 0.41, $P < 0.05$).[7] This case also demonstrates a minimally invasive sampling technique that could be considered in such scenarios. A catheter placed in the main pulmonary artery to deliver contrast for a pulmonary angiogram can also be used to aspirate tissue samples directly from the lesion. Surgical resection as part of multimodality therapy has improved survival for patients from an average of 1.5 months without treatment to approximately three years in the largest analysis of outcomes of this rare tumor.[1]

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Disclosure

No authors report any conflict of interest.

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