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CASE REPORT

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Pulmonary artery sarcoma: An important mimic of pulmonary embolism—Case reports and literature review

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Abstract

Pulmonary artery intimal sarcoma (PAIS) is a rare malignancy which closely mimics acute or chronic pulmonary thromboembolism. There are clinical and radiological characteristics which may raise suspicion of this important differential diagnosis. These include disproportionately low D-dimer, troponin T or NT-proBNP, as well as characteristic findings on CT pulmonary angiography such as the 'wall eclipsing sign' and an non-dependent position of filling defects in the large arteries. Prompt diagnosis avoids inappropriate anticoagulation and facilitates early surgical management which may improve prognosis. There is emerging evidence of an effective treatment paradigm with surgical resection and adjuvant chemotherapy. We present two cases of PAIS diagnosed at a single centre within a 2-year period. We review the literature and demonstrate the features at presentation in our cases which were suggestive of the diagnosis.

KEYWORDS

intimal sarcoma, lung embolism, lung sarcoma, pulmonary artery, pulmonary embolism

INTRODUCTION

Pulmonary artery intimal sarcoma (PAIS) is a rare high-grade malignancy of the pulmonary vasculature which closely mimics pulmonary thromboembolism (PTE).[1] With only a few hundred cases reported in the literature, a high index of suspicion is necessary to avoid delay in diagnosis and resultant poor outcomes. There are clinical and radiological characteristics which can assist in distinguishing between this malignancy and PTE. We present two cases of PAIS diagnosed at a single centre within a 2-year period with a brief review of the literature.

CASE REPORT

Case 1

A Caucasian man in his eighth decade of life presented to hospital with acute right-sided pleuritic chest pain, which had its onset during an overseas holiday. This occurred in the setting of 2 months of bilateral lower limb swelling and more recent night sweats. Upon presentation, the patient was found to be tachycardic, with a low-grade fever, but otherwise normotensive and normoxic. Troponin T was mildly elevated at 23 ng/L and no other organ dysfunction was evident on routine pathology. D-dimer and NT-proBNP were not tested at the time of presentation. Computed tomography (CT) pulmonary angiography revealed a large filling defect in the right ventricle extending into the pulmonary trunk with features of right heart strain.

Echocardiography confirmed a large, 6 cm × 4.7 cm mass occupying the right ventricular outflow tract, extending into the pulmonary trunk. The right ventricle was severely dilated with an estimated right ventricular systolic pressure of 93 mmHg. The patient underwent thrombolysis and anticoagulation via heparin infusion. Following this, the patient developed hypotension with a systolic blood pressure of 60 mmHg, which responded to minor doses of metaraminol and fluid resuscitation. The patient's respiratory failure worsened, requiring oxygen via high-flow nasal prongs. Repeat CT pulmonary angiography revealed no change

in the burden of filling defects. The patient proceeded to endovascular clot retrieval, and femoral catheters for extracorporeal membrane oxygenation were placed prophylactically prior to the procedure. Digital subtraction angiography suggested the obstruction was adherent to the right ventricular wall and despite aggressive manipulation only a small amount of the obstructive lesion could be retrieved.

Subsequent histopathology of the retrieved sample revealed necrotic tumour admixed with thrombus, with both epithelioid and sarcomatoid features. Immunohistochemical staining was positive for both cytokeratin and vimentin, suggestive of metastatic clear cell renal cell carcinoma. Positron emission tomography (PET)-CT imaging revealed intensely fluorodeoxyglucose (FDG)-avid disease in the right ventricular outflow tract extending into both the right and left main pulmonary arteries with a maximum standardized uptake value of 21. There was no evidence of extra-pulmonary disease. To reduce tumour size and relieve right ventricular strain, the patient was commenced on dexamethasone and underwent prompt external beam radiotherapy, receiving 20 Gray in five fractions.

Anticoagulation with heparin was continued due to the presence of thrombus admixed with tumour on biopsy, and bilateral lower limb deep vein thromboses were found on Doppler ultrasound. To further complicate matters, the patient developed progressive thrombocytopenia, with a platelet nadir of 33×10^9/L. Heparin-induced platelet antibody testing resulted positive, and a diagnosis of heparin-induced thrombocytopenia was made. Anticoagulation was transitioned from heparin to fondaparinux, and subsequently to rivaroxaban. The sample retrieved via endovascular catheter later resulted positive for mouse double minute 2 (MDM2) gene amplification, confirming the diagnosis of PAIS. The patient improved following radiotherapy and was discharged home.

Three months after diagnosis, the patient underwent surgical resection of the right ventricular tumour with curative intent (Figure 1: pre- and post-operative imaging). Despite some improvement following radiotherapy, the patient had a substantial burden of persistent symptoms. The patient underwent pulmonary endarterectomy of the main and bilateral pulmonary arteries, guided by cardiac magnetic resonance imaging (MRI) findings. Pulmonary valve replacement was necessary as the tumour extended through the valve into the right ventricle. Severe mitral regurgitation with a flail leaflet necessitated mitral valve replacement also. His post-operative course was largely uncomplicated. Unfortunately, repeat imaging 1 month post-operatively revealed new pulmonary nodules, measuring up to 22 mm in diameter. These increased in number and size on imaging 2 months later with evidence of local recurrence identified in the pulmonary vasculature. The patient was offered single-agent chemotherapy with doxorubicin but only tolerated a single cycle, and then was too unwell to continue. He was referred to palliative care and died 10 months after diagnosis.

Case 2

A Caucasian man in his sixth decade of life consulted his general practitioner for a 3-month history of exertional dyspnoea, with associated anorexia and 10–15 kg of weight loss over 12 months. The patient was a current smoker of tobacco, cannabis and occasionally used inhaled methamphetamine. An outpatient CT pulmonary angiogram revealed a large occlusive filling defect within the pulmonary trunk extending into the right and left main pulmonary arteries in keeping with a massive saddle pulmonary

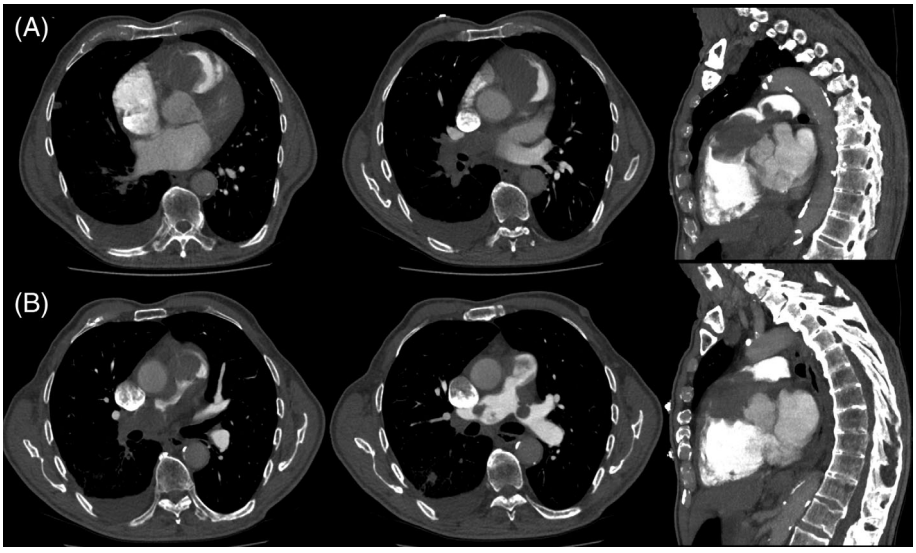

FIGURE 1 Transverse and sagittal computed tomography pulmonary angiography of Case 1. (A) Images taken at the time of diagnosis showing mass extending from pulmonary trunk into the main pulmonary arteries. Note the anterior and non-dependent position of the mass in the pulmonary trunk. (B) Images taken post-surgical resection showing debulked tumour but large volume of residual disease

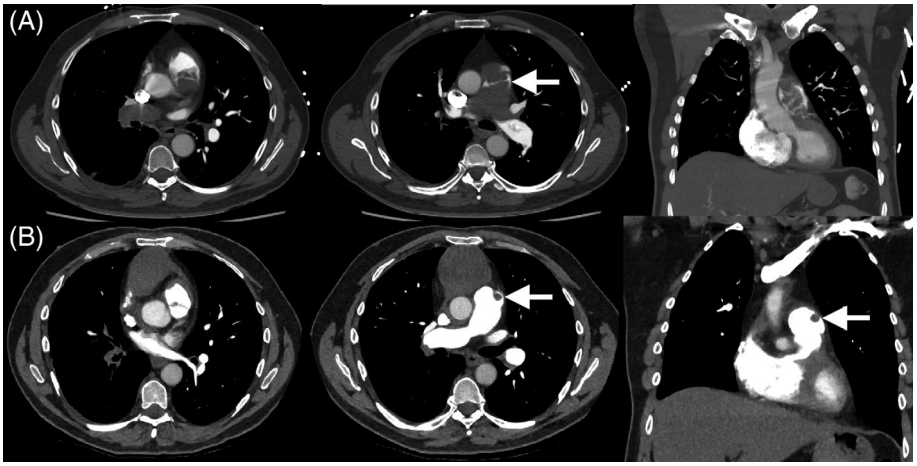

FIGURE 2 Transverse and sagittal computed tomography pulmonary angiography of Case 2. (A) Images taken at the time of diagnosis showing near-complete occlusion of both main pulmonary arteries. Again note the non-dependent position of the mass (arrow). (B) Images taken post-surgical resection showing minor residual disease in the pulmonary trunk (arrows)

embolus. There was complete occlusion of the lobar branches of the right pulmonary arterial tree, with evidence of right ventricular strain. On arrival to the emergency department, the patient was found to be tachycardic, but normotensive and normoxic. Troponin T was mildly elevated at 18 ng/L. D-dimer and NT-proBNP were not measured. Transthoracic echocardiography revealed right ventricular dysfunction and dilatation. An inferior vena cava filter was placed to prevent embolism precipitating acute decline. Reduced-dose thrombolysis was given with 50 mg of alteplase in conjunction with a heparin infusion.

Repeat CT pulmonary angiography the following day revealed no change in the burden of filling defects. A lower limb Doppler ultrasound did not reveal an underlying deep vein thrombosis. The lesions were not considered amenable to catheter-based interventions, and the decision was made to proceed with urgent open pulmonary embolectomy. The patient underwent a median sternotomy on cardiopulmonary bypass the following day. On dissection of the main pulmonary artery, a large, grey, woody tissue mass was visualized, densely adherent to the arterial wall. Frozen section histopathology during the procedure indicated that this was likely malignant tissue rather than organized thrombus, and complete excision was attempted (Figure 2: pre- and post-operative imaging). His post-operative course was unremarkable, and echocardiography revealed improved right ventricular function.

Histopathology revealed features in keeping with a PAIS, with areas of low- and high-grade tumour cells extending to all margins. The sample was positive for MDM2 gene amplification, strongly supporting this diagnosis. Post-operative CT imaging revealed persistent filling defects in segmental arteries bilaterally and mural thickening in the pulmonary trunk, with no disease evident elsewhere. Pulmonary parenchymal lesions were present, mostly in keeping with pulmonary infarction. Medical oncology recommended palliative chemotherapy given submaximal resection of the tumour. The first of six cycles of doxorubicin and ifosfamide was commenced 5 weeks post-operatively and has been well tolerated to date.

DISCUSSION

Pulmonary angiosarcomas are rare, high-grade malignant tumours originating from the pulmonary artery. They are the most common sarcoma arising from the great arteries and are sub-classified into intimal and intramural subtypes, of which the intimal subtype is far more common.[2] The first PAIS was described in the literature in 1923 with only a few hundred cases described to date.[3] It is most commonly diagnosed in the fifth and sixth decades of life, but can occur at all ages including in infancy.[4–6] There is a slight male predominance.[6–8]

Dyspnoea, syncope, chest pain, cough and haemoptysis are the most common symptoms at diagnosis.[1,6,9] Symptoms at presentation are non-specific and closely mimic those of PTE as both processes result in obstruction of blood flow from the right ventricle. Consequently, the majority of cases are initially misdiagnosed as PTE, which places patients at risk of complications from anticoagulation or thrombolysis. In several case series, the mean delay between the onset of symptoms and diagnosis was over 1 year, and diagnosis may be delayed by up to 3 years.[8,10] Our first case suffered significant haemodynamic instability and worsened hypoxia as a result of thrombolysis. The cause of this may have been due to haemodynamic changes from adherent thrombus breaking off and causing obstruction more distally in the pulmonary arterial tree.

Clinical findings which help distinguish a diagnosis of PAIS from PTE include the absence of deep vein thrombosis, a D-dimer of less than 1.0 mg/L and normal or mildly elevated cardiac biomarkers (troponin T and NT-proBNP), which are out of keeping with the burden of filling defects on CT angiography.[8] As demonstrated in our first case,

PAIS can be associated with local thrombus formation which may result in an elevated D-dimer. A similar case was reported by Sakai et al., who noted a raised D-dimer value on initial pathology, which reduced with thrombolytic therapy despite a lack of reduction in lesion size. In that case, at the time of surgical removal, no thrombus was identified, leading the authors to hypothesize that the PAIS had been surrounded by thrombus which had then been rapidly dissolved by anticoagulation therapy.[11]

A number of imaging characteristics have been identified which may assist in the diagnosis. CT angiography findings suggestive of PAIS include lobulated or polypoid filling defects, tumoural impaction, proximal lesions or heterogeneous attenuation. A finding suggested to be pathognomonic of PAIS termed as the 'wall eclipsing sign' occurs where the filling defect appears to arise from either side of the pulmonary artery.[1,8] This finding was present in 42% of a series of 26 patients with PAIS, but was not present in any of 52 controls who had PTE. In both our cases, filling defects were present anteriorly in a non-dependent position within the vessel which we propose is suggestive of PAIS rather than PTE. The presence of contrast enhancement on venous phase CT is also highly suggestive.[12] Conversely, a tubular-shaped filling defect is suggestive of PTE.[8]

PET may be useful in distinguishing between thrombus and sarcoma, with significantly higher FDG uptake in PAIS than in thrombi.[13] However, PET cannot be used to reliably exclude sarcoma as there are numerous reported cases with minimal FDG uptake.[14–16] Cardiac MRI is another available modality, which uses specific sequences to differentiate cardiac masses based on delayed retention of gadolinium within the extracellular matrix of tumours.[17] It should be noted that these additional imaging modalities are not always feasible as part of the initial diagnostic investigation in the acute setting, as in our two cases. However, PET/CT and cardiac MRI may be more reasonably considered in the investigation of subacute dyspnoea or when chronic thromboembolic pulmonary hypertension is an important differential diagnosis.

The diagnosis is most commonly made from surgical specimens after a presumed PTE has not responded to therapy as expected.[18] It is not uncommon for the diagnosis to be determined after pulmonary endarterectomy for presumed chronic thromboembolic pulmonary hypertension, with PAIS found incidentally in 1%–4% of cases.[19] A defining feature of the sarcoma is polypoid intraluminal growth with obstruction of the vessel lumen.[7] The most common location is the main pulmonary artery and at the bifurcation of the main pulmonary arteries, but they may originate in the right ventricle or more distally in the pulmonary arteries.[4] PAIS are thought to arise from intimal pluripotent mesenchymal cells, and as a result the morphological appearance and immunohistochemical characteristics of the tumour are often quite varied.[20] They tend to be myxoid with pleomorphic areas and may show varying degrees of differentiation. This can make histopathological diagnosis difficult as seen in our first case. The presence of the MDM2 mutation strongly supports the diagnosis.[21,22] Case series report the presence of metastatic disease at diagnosis in approximately 30%–50% of patients, with the majority confined to the lung.[8,23] Recurrence at distant sites after surgery suggests a higher rate of undiagnosed metastatic disease.[24] Metastatic disease at diagnosis is strongly associated with reduced survival.[25] Local recurrence appears to occur in approximately 50% of cases after resection, and distant metastasis in approximately 20%–30%.[23,24]

Surgery has been utilized as the primary management option for primary tumours, local recurrences and metastatic lesions. The goal of surgical intervention is cure; however, this is not commonly achieved, with only a few reports of complete resection without recurrence reported in the literature.[13,26,27] Pulmonary endarterectomy is the most common procedure, with graft reconstruction of the pulmonary artery if required. There is evidence that pulmonary endarterectomy provides better symptomatic and haemodynamic outcomes than more limited tumour resection, with a trend towards improved survival.[24] Pneumonectomy is sometimes performed for the rare case of unilateral disease, and is associated with trend towards improved survival.[28] Intra-arterial stents are rarely used, there is a case report of successful use of a stent as bridge to pneumonectomy; however, another resulted in death during placement.[29,30] There are two cases of heart–lung transplantation reported in the literature, but unfortunately both recipients died at 5 months.[31–33] Our two cases improved symptomatically following surgery; however, the benefit was short lived for our first case and neither case remained disease-free.

A multi-modal approach with adjuvant chemotherapy may contribute to improved life expectancy; however, the role of radiotherapy remains unclear.[2,13,34] Radiotherapy was symptomatically beneficial in our first case; however, the effect on long-term survival is unknown. First-line chemotherapy is doxorubicin with or without ifosfamide or cisplatin, based on the established treatment paradigms for metastatic soft tissue sarcoma. There are case reports of response to vinorelbine as second-line treatment, and there are reports of prolonged response from chemoradiotherapy alone.[34–36] There is emerging evidence that small molecule tyrosine kinase inhibitors may be effective in the palliative management of PAIS. The multi-target tyrosine kinase inhibitor pazopanib has been shown to prolong survival in metastatic soft tissue sarcoma and to suppress the proliferation of PAIS in tissue culture.[22,37] There is a single case report describing clinical response of PAIS to pazopanib.[27]

The prognosis of PAIS is usually poor due to the difficulty in affecting complete surgical resection, early embolic dissemination throughout the lungs and resistance to both chemotherapy and radiotherapy. Prognosis strongly depends on the location of the primary tumour and the presence of metastatic disease with reported median survival rates varying between 7 and 26 months.[9,13] Three retrospective studies of patients with surgical resection followed by systemic chemotherapy reported 2-year survival rates between 35% and 45%.[9,13,32]

PAIS is a rare differential of acute or chronic PTE, and our cases demonstrate that a high degree of suspicion is

required in order to make a timely diagnosis and achieve optimal outcomes. There is evidence of several clinical and radiological features which raise the suspicion of the diagnosis. Whilst it is unreasonable to expect PAIS masquerading as acute pulmonary embolism to be diagnosed prior to anticoagulation or thrombolysis, it is hoped that recognition of suspicious features will result in investigation for this rare differential in the follow-up period after presentation, or during the evaluation of chronic thromboembolic pulmonary hypertension. Evidence is emerging of an effective treatment paradigm once a diagnosis is made.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

Colin Tuft is the primary author and reviewed the literature. Krishan Maheepala is the secondary author reviewed the literature. Steven Lindstrom and Ajantha Raguparan are the supervising physicians and edited the manuscript. Anas Naeem provided intensive care input. Suhrid Lodh provided interventional radiology input.

DATA AVAILABILITY STATEMENT

Data available on request due to privacy/ethical restrictions.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this case report and accompanying images.

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