CASE REPORTS

A RARE CASE OF PULMONARY ARTERY SARCOMA

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INTRODUCTION

Pulmonary artery sarcoma (PAS) is a rare and aggressive mesenchymal tumor with an overall poor prognosis[1-5]. Due to similar clinical and radiologic findings, PAS is often misdiagnosed as a pulmonary embolism (PE) frequently leading to prolonged anticoagulation therapy, which delays the correct diagnosis [1-3]. By presenting this clinical case our objective is to emphasize characteristic CT findings that favour a neoplastic origin of a pulmonary intravascular filling defect. PET-CT and MRI have also an important potential role in its diagnosis and therapeutical management[1-3].

CLINICAL CASE

A 48-year-old man, known smoker with hypertension, was referred to the ER with a history of progressive shortening of breath and dry cough for the past 2 months, aggravated in the last week. There was no history of chest pain, palpitations or syncope. Laboratory findings showed a slightly elevated D-dimer and ECG showed a S1Q3T3 pattern, raising the possibility of PE. Patient was hemodynamically stable, apiretic, without hypoxemia or clinical signs of deep venous thrombosis. Chest film showed an increase of cardiothoracic ratio suggesting cardiomegaly with no other relevant findings (Figure 1).

Complementary echocardiogram showed right chamber enlargement, with an estimated pulmonary artery systolic pressure of 80 mmHg. A turbulent flux was noted in the main pulmonary artery, with suggestion of a mass attached to it.

A contrast-enhanced CT (CECT) was performed revealing exuberant hypodense filling defects within the lumen of the pulmonary trunk and main pulmonary arteries,with proximal bulging extension to the right ventricular outflow tract (Figure 2). The filling defects were continuous, mainly in a peripheral position, adjacent to the vessel wall, with contrast flowing in the center of the vessels, resembling chronic PE. However, the lesions had a mild expansive effect, some areas suggesting extraparietal extension, subtle spiculation and mild late post contrast enhancement. These findings were not typical of PE, which raised concern for other diagnosis, in particular malignancy.

To exclude this possibility, a PET-CTscan was requested, revealing high F-18 FDG-uptake (SUVmax 8) along the arterial walls of the pulmonary arteries (Figure 3), corresponding to the filling defects observed on CT, confirming the suspicion of a malignant disease.

A surgical biopsy was performed, with the histological diagnosis of a pulmonary artery undifferentiated pleomorphic sarcoma. The infiltrating mass was not resectable and the patient was referred for chemotherapy, having survived only 4 months after diagnosis.

DISCUSSION

PAS is a very rare malignancy, arising from the intimal layer of the pulmonary trunk or main arteries 1-5. Retrograde extension to the right ventricular outflow tract or pulmonary valve has also been reported3. It generally presents in middle-aged adults, although the age of diagnosis is broad [2]. There is a slight female predominance[2,4]. Presenting symptoms may include dyspnea, cough, chest pain and progressive right heart failure. Laboratory tests are often nonspecific [2,3].

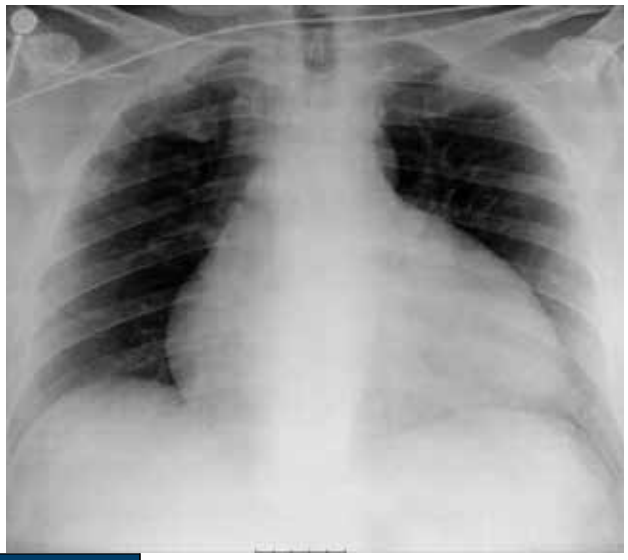

Figure 1 *Cardiomegaly on chest film.*

PAS is frequently misdiagnosed as a PE and this is explained by its similarities in terms of clinical features and imaging findings [1-5]. In a CECT examination, the presence of a low-attenuation filling defect occupying the lumen of the main or central pulmonary arteries is a typical finding in PE. Although incredibly rare compared to PE, PAS may also manifest with a pulmonary filling defect in CECT, many times being misdiagnosed as a PE[1-3].

Findings that should alert the radiologist to the possibility of PAS other than PE include:expansion of the pulmonary wall, bulging proximal end of the tumour, extraluminal tumoral extension or late contrast enhancement, findings not present in PE[1,3]. Additionally, in PAS, the proximal part of the filling defect is usually prominent and lobulated, different from the typical straight or obtuse margins of PE thrombus[3]. Furthermore, lack of improvement despite adequate anticoagulation, no evidence of deep venous thrombosis and the presence of distant metastasis corroborate this diagnosis[1].

Both PET-CT and MRI can be useful, showing FDG-uptake or heterogeneous enhancement, supporting the diagnosis of a malignant lesion rather than a blood thrombus, which shows no FDG-uptake or enhancement[1-3].

Despite the recent advances in imaging modalities, definitive diagnosis requires histopathologic characterization. Because tumor cells are present in the vascular intima, endovascular catheter biopsy can directly approach a lesion in the vascular lumen, being a reasonable diagnostic method5. In our clinical case,

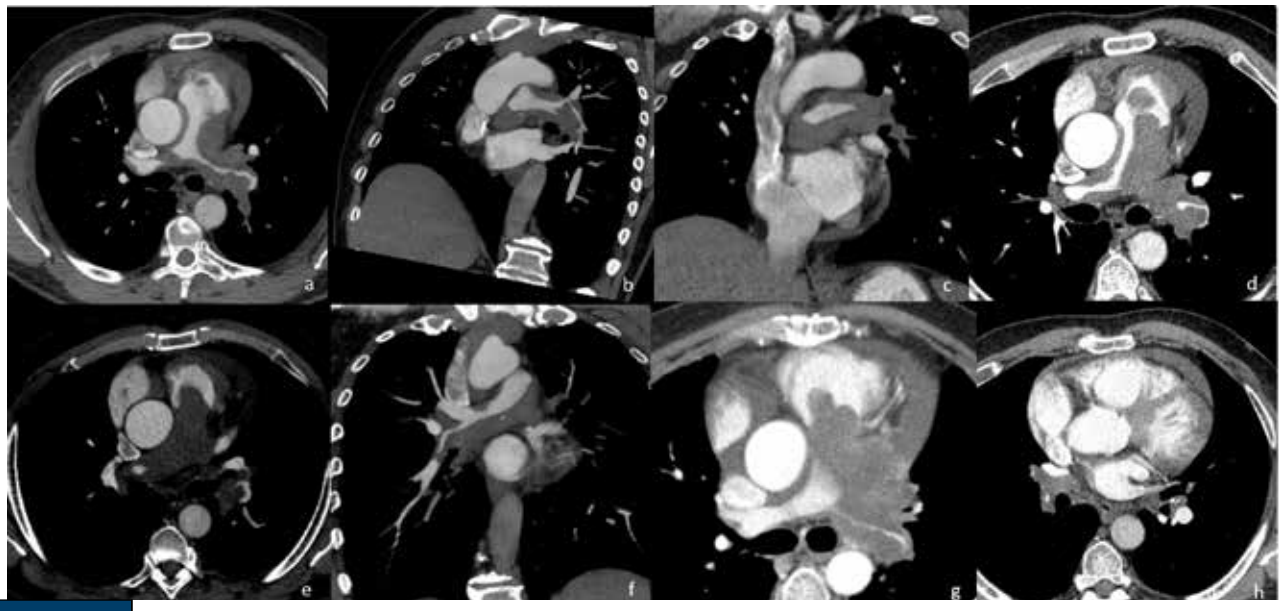

Figure 2 *CECT (a-h) showing low density filling defects occupying and expanding the trunk and main pulmonary arteries.*

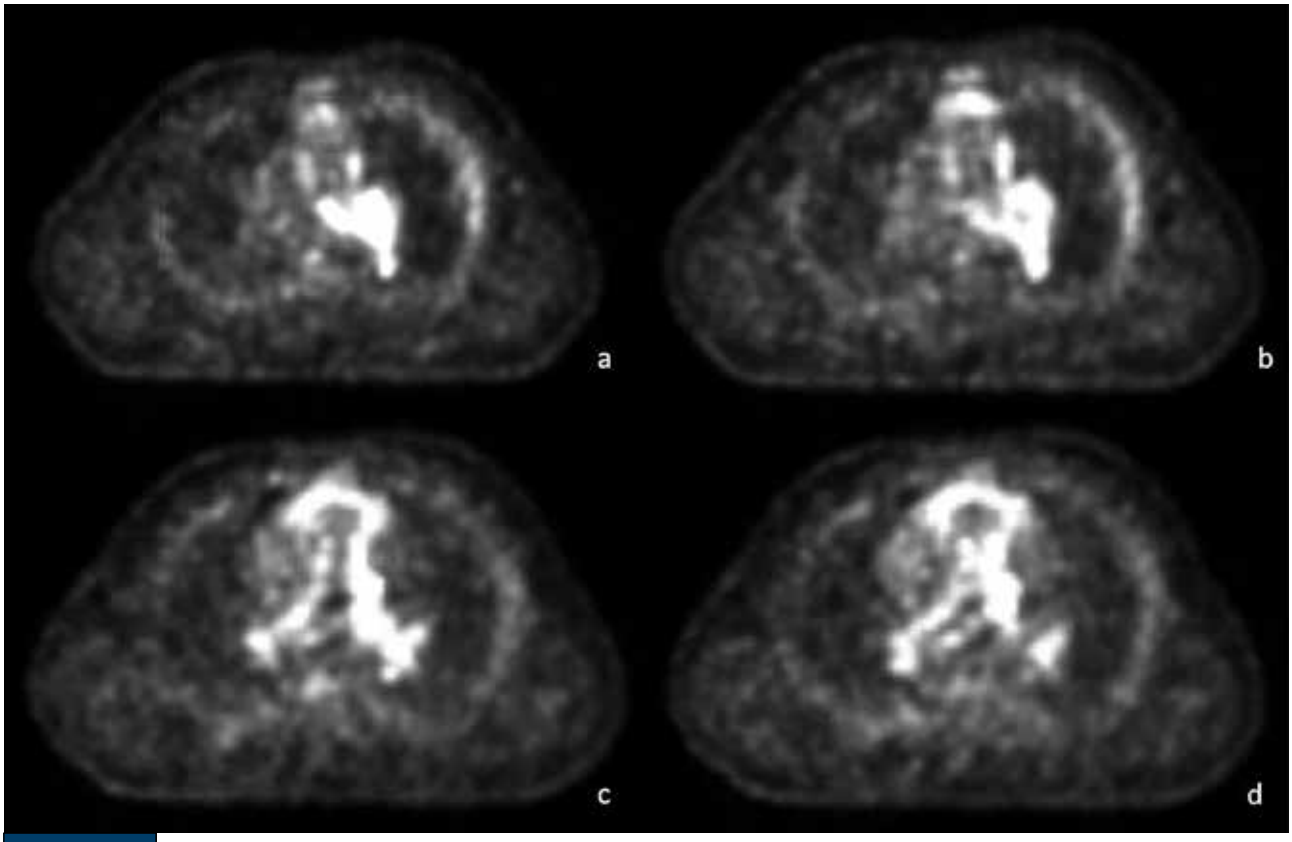

Figure 3 *PET-CT scan (a-d) showed correspondent high uptake.*

with the prospect of characterization and possibility of surgical excision, the cardiovascular surgeons chose to select surgical biopsy.

The prognosis of PAS is related to local invasion and metastization, with a poor median survival of only few months in untreated patients[2]. The only chance of a potential cure consists, when possible, in radical surgical resection[4,5]. The best results are obtained through pulmonary arteriotomy with resectionof the tumor and its implantation site, followed by reconstruction of the main pulmonary artery with a pericardium homograft[4]. Distal embolectomy of metastasized PAS emboli in segmental pulmonary arteries may further extend survival for these patients[4]. The role of chemotherapy and radiation remains unclear, although it seems to improve survival rate, especially after surgery[3,4].

CONFLICT OF INTEREST

The authors declare no conflict of interests.

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