# Pulmonary artery osteosarcoma masquerading as pulmonary thromboembolism: the role of multimodality imaging

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## Abstract

Primary pulmonary artery sarcoma (PAS) is an extremely rare malignant disorder that presents like pulmonary thromboembolism (PE). Primary osteogenic sarcoma in the pulmonary artery (PA) is even rarer and can produce osteoid or cartilaginous matrix. Few studies have described the radiographic characteristics of osteosarcoma of the PA. We there report a case of a 78-year-old male patient with osteosarcoma in the PA where the patient went through surgical treatment after careful multimodalityimaging assessment. The patient was admitted to our hospital with the nonspecific symptom of heart failure. Multimodality imaging showed the primary lesion adhering to the arterial wall but without invading into surrounding tissues. PET/CT showed signs of hypometabolic activity within the lumen of the main PA. Cardiac MRI showed preserved left ventricular systolic function. CT showed distinctive features of PA osteosarcoma (a slightly hyperdense mass with calcification in pulmonary trunk).

Keywords Pulmonary artery; Osteosarcoma; Dyspnoea; Magnetic resonance imaging; Computed tomography

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### Introduction

Pulmonary artery osteosarcoma, a subtype of pulmonary artery sarcoma (PAS), has been rarely reported, with a prevalence of about 4% in the literature.<sup>1</sup> Here, we present a case of a 78-year-old male patient admitted to our hospital due to progressive heart failure symptoms.

# **Case Report**

A previously healthy 78-year-old male patient presented with progressive dyspnoea on exertion (New York Heart Association Class III), wheezing, and dry cough of 3 months duration. His physical examination demonstrated unremarkable results. Heart sounds were normal on cardiac auscultation except for a loud pulmonic component (P2) of the second heart sound. The majority of biochemical assays including BNP were normal, except for mildly increased fasting blood glucose concentration (6.40 mmol/mL, reference values <6.05) and slightly increased CA-19-9 in the serum (32.86 U/mL, reference values <31.3). The electrocardiography was normal. Transthoracic echocardiography at local hospital showed left ventricular mild diastolic dysfunction. Pulmonary thromboembolism (PTE) was highly suspected according to subsequent contrast-enhanced chest computed tomography (CT) and PET/CT scan. After a 2 month anticoagulation therapy, the symptoms did not relieve, and the patient was readmitted to our hospital. Our CT scan (Figure 1) depicted a large filling defect  $(8.5 \times 3 \text{ cm})$  in the main pulmonary artery with polyploid ossification, patchy ground-glass opacification in the right middle and lower lobes. Cardiac MRI revealed right heart dilatation, left ventricular ejection fraction of 59%, right ventricular ejection fraction of 41%, and T2-weighted dark blood sequence

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Figure 1 Contrast-enhanced chest computed tomography scan.



Figure 2 Dark-blood T2-weighted cardiac MR scan.



Figure 3 Steady-state free-precession (SSFP) cine cardiac MR scan.



showed a slightly hyperintense soft tissue-filled mass (*Figure 2*). Steady-state free precession cine demonstrated a mass with beaded periphery distal to the pulmonic

Figure 4 Delay enhancement cardiac MR scan by phase-sensitive inversion recovery (PSIR) sequence.



Figure 5 Whole-body positron emission tomography (PET/CT).



valve, moving with cardiac cycle and obstructing pulmonary trunk (*Figure 3*, Supporting Information, *Video S1*). Post-gadolinium delayed enhancement sequence showed peripheral hyperenhancement consistent with neoplasm (*Figure 4*). PET/CT showed hypometabolic activity in the pulmonary trunk without metastases in other organs, such as the liver and the lungs (*Figure 5*). Pulmonary angiography further revealed neovascularity in the mass (*Figure 6*, Supporting Information, *Video S2*). The patient underwent surgery with pulmonary endarterectomy through median sternotomy and cardiopulmonary bypass. Pathologic examination revealed a pulmonary artery low-grade osteosarcoma.

#### Figure 6 Pulmonary angiography.



#### Discussion

Different histological subtypes of pulmonary artery sarcoma (PAS) have been reported in the literature. To our knowledge, the current case of pulmonary artery osteosarcoma has not been reported in detail.<sup>2</sup> We found several imaging features of this subtype PAS, including ossification within the mass, acute angles between mass and the vessel wall, hypometabolic activity of FDG-PET scan, and absence or occurrence of distant metastasis, all consistent with the low-grade osteosarcoma. The clinical presentation of PAS is usually nonspecific,<sup>3</sup> and its radiologic appearances often lead to the misdiagnosis of PTE.<sup>4</sup> However, the treatments for both diseases are quite different. An accurate and timely diagnosis is often challenging. Aggressive surgery resection is

the primary treatment for the localized lesion. The pathophysiology of PAS is unclear, but many predisposing risk factors are linked to the occurrence of PAS.<sup>5</sup> In this case, the pathogenic factor may be his professional exposure to vinyl chloride.<sup>6</sup> In the diagnosis of pulmonary artery osteosarcoma, multimodality imaging should be underscored to avoid inappropriate diagnosis and a delayed intervention. Clinicians should be aware of the key clinical and radiographic characteristics that differentiate this disease from PTE, because timely diagnosis coupled with surgical resection can improve the prognosis of the disease.

#### **Conflict of interest**

None declared.

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### Supporting information

Additional supporting information may be found online in the Supporting Information section at the end of the article.

Video S1. Supporting Information. Video S2. Supporting Information.

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