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Case Report

Primary Intimal Sarcoma of the Pulmonary Artery in a 25-Year-Old Woman with Dyspnea and Palpitation: A Case Report

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Keywords
Intimal sarcoma · Chondroblastic osteosarcomatous differentiation · Pulmonary artery · Pulmonary embolism

Abstract
Intimal sarcoma arising from the tunica intima of both systemic and pulmonary circulations is a rare disorder, whereas intimal sarcoma with chondroblastic osteosarcomatous differentiation (ISCOS) is even rarer. We present the case of a 25-year-old woman with ISCOS of the pulmonary artery (PA) where the patient went through surgical treatment after careful imaging assessment under a rather emergent situation. A 25-year-old Japanese female presented to our hospital with the chief complaints of dyspnea and palpitations on exertion. Upon arrival, she had systolic murmur, moderate tricuspid regurgitation, and possible pulmonary hypertension. A contrast-enhanced chest computed tomography (CT) showed dilatation of the main PA, filled with a hypodense area with calcification adjacent to the right and left PA. The calcified lesions within the tumor were the key findings suggesting osteoid-forming sarcoma, differentiating it from pulmonary embolism. Due to presence of critical symptomatic obliteration of the pulmonary circulation, an emergency surgery was performed. A whitish shiny mass filled the lumens from the main PA to the bilateral main PAs. The tumor was not attached to the surrounding intima, except for a slight attachment to the left interlobar PA, and could be completely removed from the vessel lumen. Based on the pathological findings, it was diagnosed as a primary ISCOS of the PA, which correlated with the findings of the CT, namely

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intratumoral calcification. Although the diagnosis-making is quite challenging, multidisciplinary collaboration between clinicians, radiologists, and pathologists is crucial for reaching the correct diagnosis.

Introduction

Intimal sarcoma (IS) of large blood vessels is a rare malignant disorder, arising from the tunica intima of both systemic and pulmonary circulations, including the aorta and/or the pulmonary arteries (PA) [1, 2]. The median age at the time of diagnosis of IS depends on the sites of origin: 48, 50, and 62 years for pulmonary vessels, smaller systemic vessels, and larger systemic vessels, respectively. It occurs more frequently in females with a male to female ratio of 7:10 [1, 3, 4]. Patient outcome of this disorder is very poor, as previously reported, with a mean survival time of patients with pulmonary and aortic IS of 13–18 and 5–9 months, respectively [1, 2]. IS shows various histological subtypes of differentiation, such as fibrosarcoma, leiomyosarcoma, anaplastic sarcoma, osteosarcoma, and chondrosarcoma [5]. The standard therapy currently available for IS is only surgical resection, as no systemic treatment has been established for this rare disease [2, 6, 7].

Whereas IS of the PA is rare, IS with chondroblastic osteosarcomatous differentiation (ISCOS) arising from the PA is even rarer as, to the best of our knowledge, only 15 cases have been reported [8, 9]. There are numerous challenges in making the diagnosis of sarcoma of the PA because its clinical features may resemble those of nontumorous disorders involving the PA, such as pulmonary embolism or other PA tumors [9]. Clinicians may initiate anticoagulant therapy for PA tumor and pulmonary thromboembolism before a correct diagnosis is made, as previously reported [8–11]. Tavora et al. [9] reported that the initial diagnosis of two-thirds of cases with ISCOS of the PA was pulmonary embolism. Sarcomas of the PA generally cause similar symptoms regardless of histological type, thus a definitive diagnosis would be possible only by surgical specimen or postmortem examination.

We herein report the rare case of a 25-year-old woman who presented with dyspnea and palpitation and was diagnosed with ISCOS of the PA by means of surgical treatment. The calcified lesions within the tumor, possibly due to osteosarcomatous differentiation, were the key findings on computed tomography (CT) to discriminate our case from pulmonary embolism.

Case Presentation

A 25-year-old Japanese female presented to our emergency department with the chief complaints of dyspnea and palpitations on exertion, starting 1 month ago. Upon arrival, physical examination revealed systolic murmur. The bedside ultrasound examination demonstrated moderate tricuspid regurgitation and possible pulmonary hypertension and the patient was hospitalized. A contrast-enhanced chest CT showed dilatation of the main PA, filled with a hypodense area with calcification adjacent to the right and left PA. The lumens of the main PA and the hilar areas of the right and left PA appeared almost obliterated by the mass; however, the mass was not attached to the pulmonary valve and did not extend into the peripheral parts of the right and left PA (Fig. 1). The differential diagnosis included primary PA tumor and pulmonary thromboembolism, but we suspected it to be a PA tumor based on the radiological findings: a relatively poor contrast effect on the lesion with calcification. Lung perfusion scintigraphy revealed decreased blood flow in the whole bilateral lungs, except for

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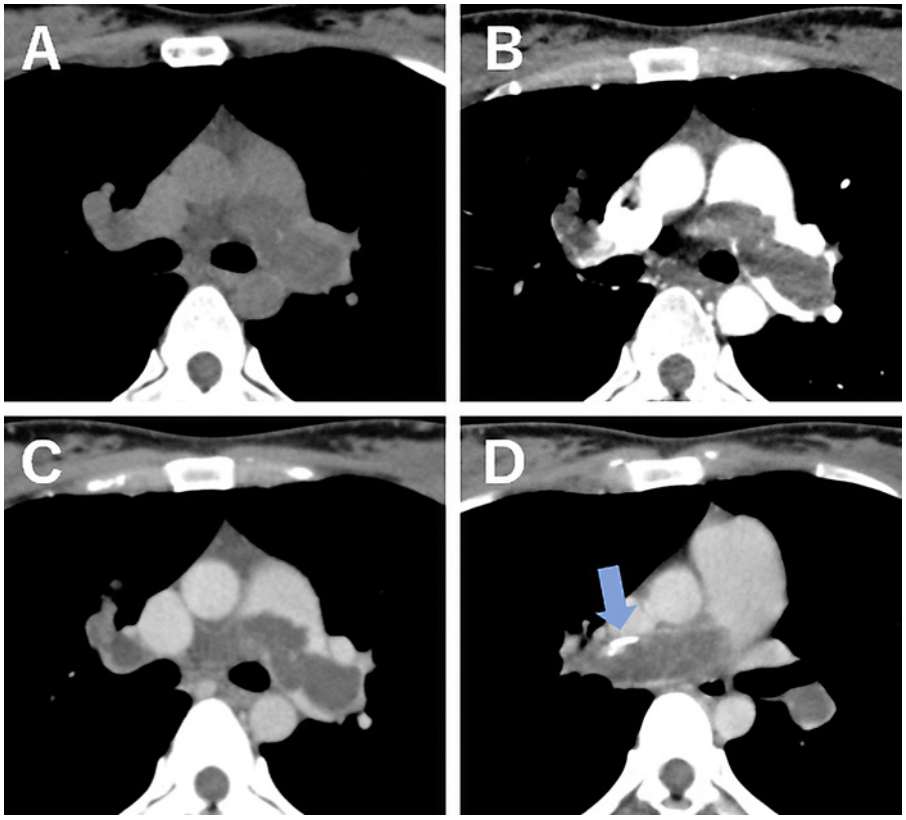

Fig. 1. A contrast-enhanced chest CT scan of the patient showed a poorly marginated mass within the pulmonary arteries. **A** Raw CT scan. **B** Arterial phase. **C**, **D** Venous and delayed phases. A periphery of the lesion contained a more hyperdense area, and calcification was observed within the tumor (blue arrow, **D**).

the left lung upper lobe. Due to critical symptomatic obliteration of the pulmonary circulation, an emergency surgery was performed on the second day of hospitalization. Preoperative FDP D-dimer was 1.9 μg/mL, slightly higher than the normal limit (within 1 μg/mL).

Following a median sternotomy and institution of cardiopulmonary bypass, deep hypothermic circulatory arrest was induced for the removal of the tumor. The longitudinal incision was made on the main PA extending into the left PA (Fig. 2A). A whitish shiny mass filled the lumens without any attachment to the surrounding intima, except that the tumor was attached to the intima of the left interlobar PA. The tumor was completely removed from the vessel lumen (Fig. 2B). Next, the longitudinal incision of the right PA behind the aorta and the superior vena cava was extended to the right interlobar PA. The neoplasm had no attachment to the intima in this area and was obliterated by the segmental branches of the right PA. The tumor was extracted and completely removed from the vessels, and the peripheral ends of the tumor demonstrated a finger-like appearance (Fig. 2C). After complete removal of the tumor and copious irrigations, the incisions were simply closed using 6-0 polypropylene sutures. The postoperative CT scan confirmed that no tumorous mass was left behind in the PAs.

Gross pathology showed a soft-to-hard whitish-brown tumor. Microscopically, spindle cells with marked cytological atypia proliferated with tumor osteoid formation. There were also lobular proliferations of chondroid islands composed of atypical chondroblasts (Fig. 3).

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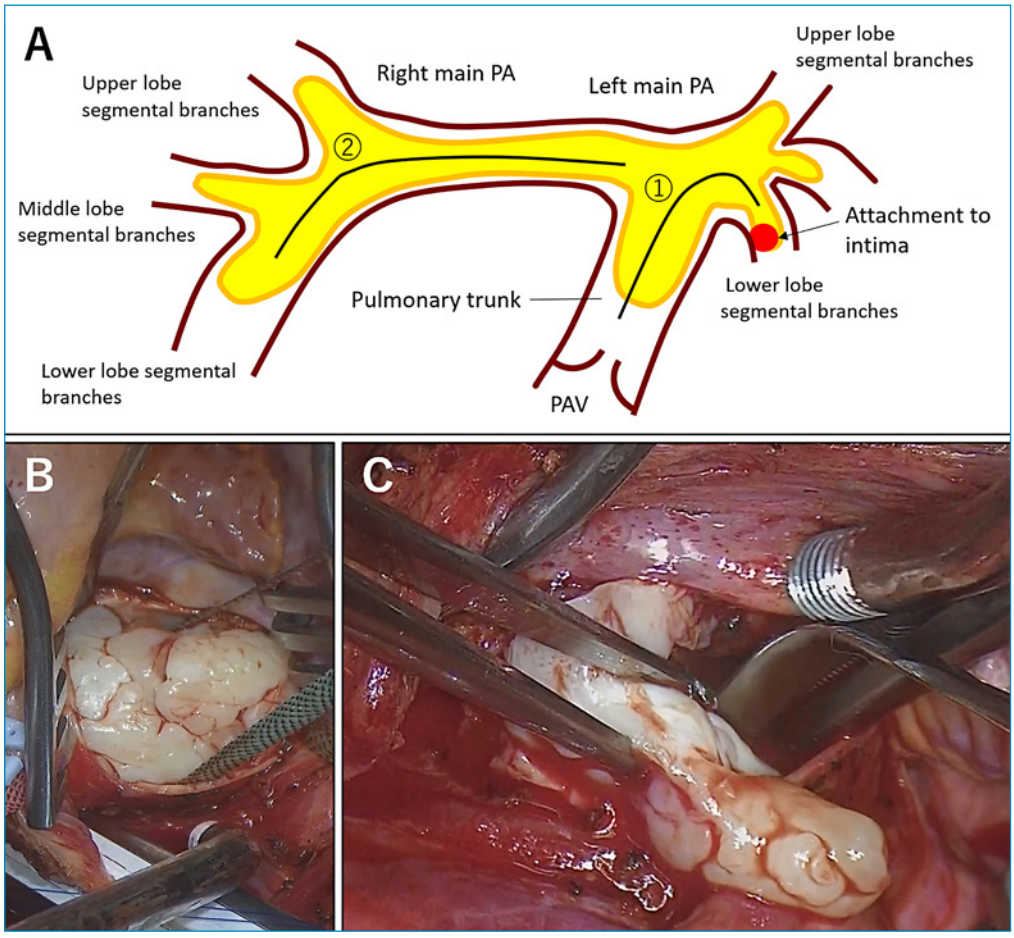

Fig. 2. Intraoperative findings of the tumor. **A** Location of the tumor, highlighted in yellow, is shown. 1 and 2 indicate incision lines of the artery. **B** The tumorous lesion occupied nearly 90% of the lumen of the pulmonary trunk. **C** A whitish shiny tumor filling the lumen of the right PA with a round finger-like structure at the edge.

Based on the pathological findings as well as the results of the clinical examination that there was no possible primary tumor, it was diagnosed as a primary ISCOS of the PA.

She received adjuvant chemotherapy, but 5 months later, a contrast-enhanced chest CT scan showed a hyperdense lesion with calcification at the upper hilum of the right lung, indicating the recurrence of the disease. Right upper lobectomy was performed, and the resected specimen contained a neoplastic lesion with similar pathological features to the primary lesion. Intraoperatively, a pleural metastatic nodule was also found and resected, which was of the same pathological characteristics. The patient is currently being followed up in an outpatient clinic without any known complications 16 months after the initial surgery.

Discussion and Conclusion

We experienced a quite rare case of primary ISCOS of the PA. With careful imaging assessment, we managed to make a correct preoperative diagnosis. Should we have first diagnosed her condition as thromboembolic disease of the PA and initiated the anticoagulant

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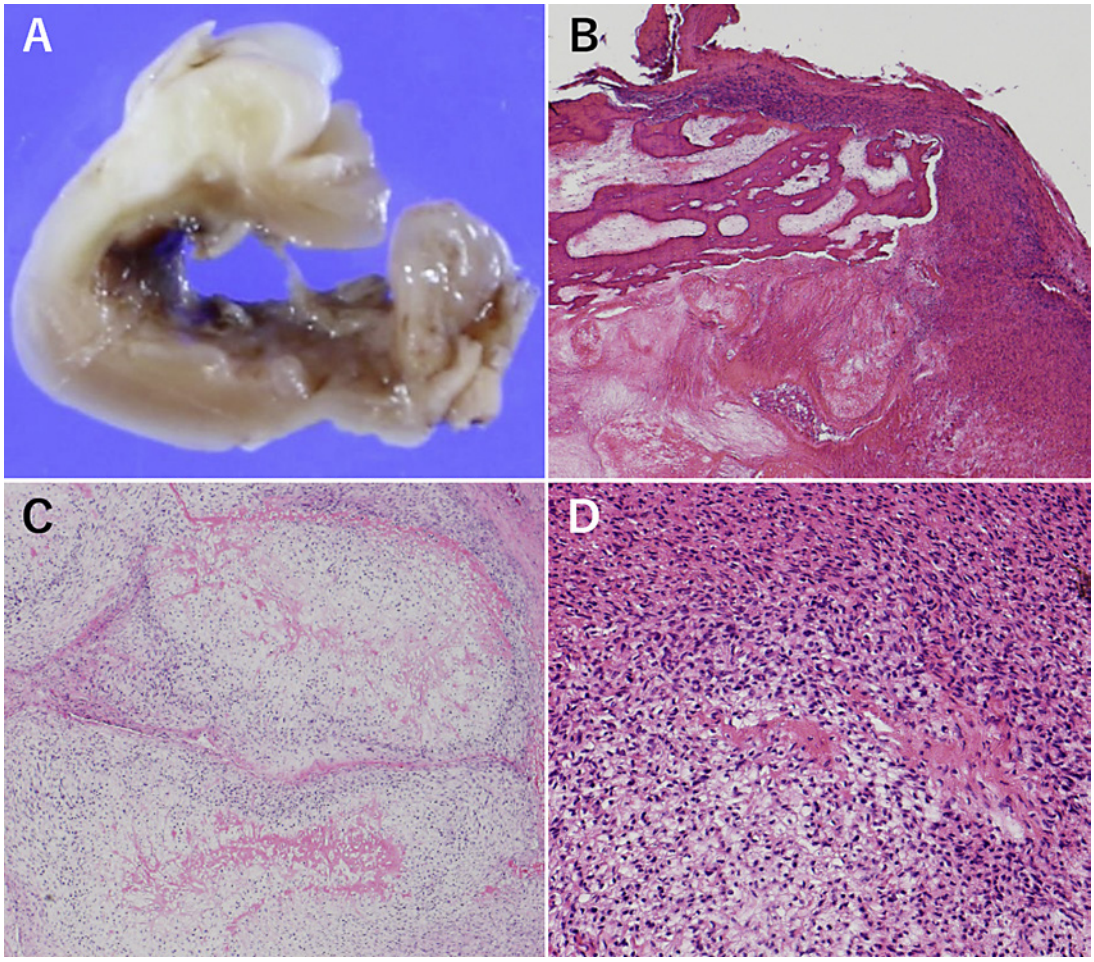

Fig. 3. Intimal sarcoma of chondroblastic osteosarcomatous differentiation. **A** Gross picture of shiny, brownish-white lesion attached to the wall of the pulmonary artery. **B** Microscopic picture of the tumor, showing proliferation of spindle cells with tumor osteoid formation. **C** In fibrous and myxoid background, spindle cells with atypia proliferate in a fascicular growth pattern with tumor osteoid formation. **D** Proliferation of spindle cells with cytological atypia with formation of chondroid matrix. Eight mitotic figures were observed in 10 high-power fields.

therapy, this would have been lethal, losing the appropriate timing for surgery. In our case, the chondroblastic osteosarcomatous differentiation was clearly associated with the calcified lesions within the tumor detectable on CT (Fig. 1D). Among 4 cases, including the present one, recorded as primary IS of the PA in our institution (Table 1), another case (case 1) having the feature of ISCOS showed a similar calcified lesion on CT (data not shown). Although calcification could even occur within thrombi in chronic pulmonary thromboembolism [12], it would be an important finding suggesting osteoid-forming sarcoma, including ISCOS. A contrast-enhanced magnetic resonance imaging (MRI) might be useful, though MRI scan was not performed due to an emergent situation.

IS originating from the PA commonly metastasizes to the lungs as it did in our case. A review of the literature showed 23 cases, of which 15 cases (65%) developed lung metastasis. Other metastasized sites were the pleura, brain, bone, liver, and others with the percentages of 9, 17, 17, 9, and 17%, respectively [2, 6, 13, 14]. Such information, especially concerning the brain, might be useful for careful follow-up of patients with IS.

Table 1. Four cases of primary intimal sarcoma of the pulmonary artery diagnosed at our institution

Case	Sex	Age, years	Histological subtype	Chief complaint	Adjuvant treatment	Outcome
1	Female	64	Osteosarcoma	Chest pain	None	NED (4 months)
2	Female	75	Angiosarcoma	Bloody sputum	None	Death with recurrence (SVC) in 8 months
3	Female	77	Leiomyosarcoma	Dyspnea	RT	NED (8 months)
4*	Female	26	Osteosarcoma	Dyspnea	CT	Alive with recurrence (lung) in 16 months

* Current case. NED, no evidence of disease; SVC, superior vena cava; RT, radiation therapy; CT, chemotherapy.

We have experienced a rather rare case with ISCOS. Although the diagnosis-making is quite challenging and can be urgent with regard to its location, multidisciplinary collaboration between clinicians, radiologists, and pathologists is fundamental to reach the correct diagnosis.

Statement of Ethics

Written informed consent for the publication of the present case report and any accompanying images was obtained from the patient.

Conflict of Interest Statement

The authors declare that they have no conflicts of interest to disclose.

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Author Contributions

H.O., A.A., and T.S. conducted the histological assessment. T.Ar. conducted the presurgical imaging assessment. T.As performed the surgery and provided the clinical information. H.O., Y.H., and H.T. wrote the manuscript. T.As. and T.S. reviewed and edited the manuscript. All authors contributed to discussions and agreed with the final version of the submitted manuscript.

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