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CARDIOVASCULAR FLASHLIGHT
Cardiac imaging

A rare case of intimal sarcoma mimicking pulmonary stenosis

Keiko Sumimoto , Yu Taniguchi *, Noriaki Emoto , and Ken-Ichi Hirata

Division of Cardiovascular Medicine, Department of Internal Medicine, Kobe University Graduate School of Medicine, 7-5-2, Kusunoki-cho, Chuo-ku, Kobe 650-0017, Japan

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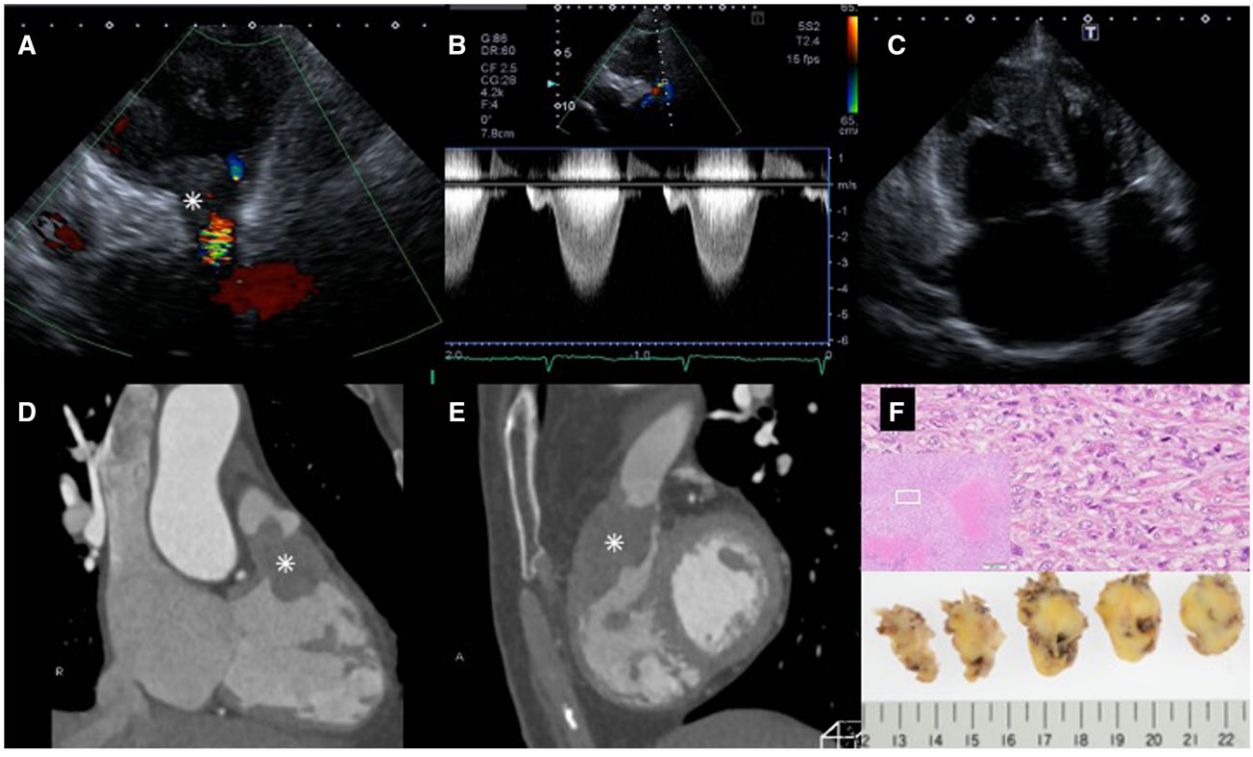
A 73-year-old woman with progressive dyspnoea and recurrent presyncope for 4 months was suspected of having severe pulmonary valve stenosis and was referred to our hospital for treatment. Transthoracic echocardiography revealed a large space-occupying mass extending from the right ventricular outflow to the main pulmonary artery, infiltrating the pulmonary valve (*Panel A*; *Video 1*). The mass caused severe pulmonary stenosis with a peak gradient of 72 mmHg and a peak velocity of 4.2 m/s (*Panel B*). It also showed right ventricular dilatation (*Panel C*), hypertrophy, and severe tricuspid regurgitation. Cardiac computed tomography confirmed an irregular mass nearly occluding the main pulmonary artery (*Panels D* and *F*, *Video 2*).



* Corresponding author. Tel: +81 78 382 5846, Fax: +81 78 382 5859, Email: yu.taniguchi007@gmail.com
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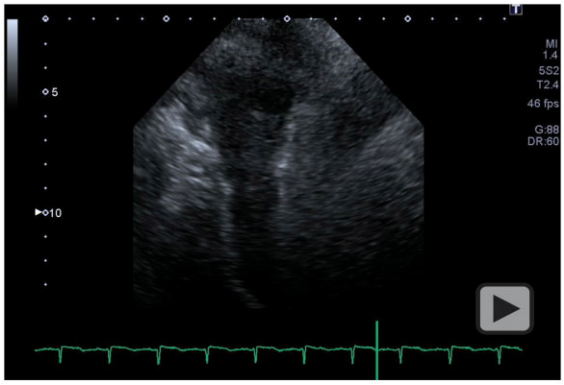
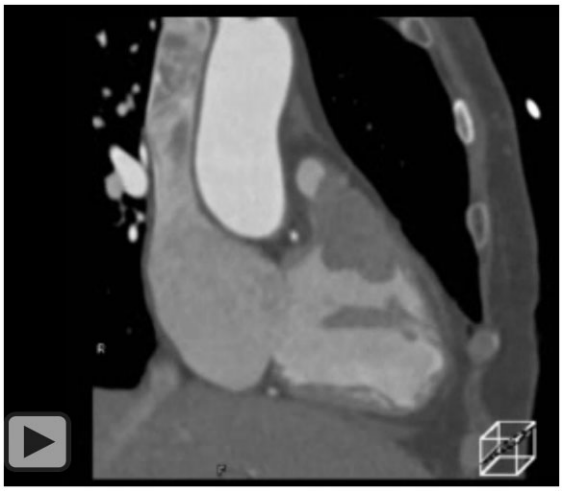


Video 1 Transthoracic echocardiography showing a mass extending from the right ventricle to the main pulmonary artery, infiltrating the pulmonary valve.



Video 2 Cardiac computed tomography showing a large space-occupying mass extending from the right ventricle to the main pulmonary artery, nearly occluding the pulmonary artery.

On the basis of her presentation and imaging findings, an emergency operation was scheduled. Intraoperative histologic examination revealed that the mass was a sarcoma (*Panel F*). As intimal sarcoma of the pulmonary artery has a very poor prognosis even with treatment, we decided to perform minimally invasive tumour resection but not complete resection. The tumour was resected by pneumonectomy and reconstruction of the right ventricular outflow tract, including pulmonary valve replacement. Adjuvant chemotherapy was initiated subsequently. One year post-operatively, she had no tumour recurrence.

Pulmonary artery intimal sarcoma involving the pulmonary valve is a very rare disease. The diagnosis is often challenging. Early diagnosis may allow better management of this disease. In this case, careful observation of the pulmonary artery through transthoracic echocardiography was the first step towards diagnosis.

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, was obtained from the patient in line with COPE guidance.

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