

# Coronary intervention for severe stenosis in the ostial right coronary artery with cardiac angiosarcoma: a case report

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## Background

Although rare, angiosarcoma is the most common type of cardiac primary malignancy. This disease can cause life-threatening complications and the prognosis remains poor. There is no standard approach to care, and clinical judgement is exercised on a case-by-case basis. Tumour progression causes serious complications, such as heart failure and vascular disruption.

## Case summary

A 64-year-old Japanese woman presenting with a right atrial tumour was referred to our department. Tumour biopsy revealed that the patient suffered from angiosarcoma. We performed a lumpectomy to excise the tumour, but due to tissue adhesions in and around the right atrium, the malignancy could not be completely removed. After 3 years of chemotherapy, the patient was admitted to our hospital with increased chest pain. Emergency coronary angiogram revealed severe stenosis of the ostial right coronary artery. Intravascular ultrasound (IVUS) and computed tomography suggested coronary compression due to cardiac angiosarcoma. In this study, we report a unique case of advanced cardiac angiosarcoma, presenting as unstable angina, which was successfully treated with percutaneous coronary intervention using stent implantation.

## Discussion

Due to the rarity of cardiac primary angiosarcoma, many symptoms are misdiagnosed until mechanical complications arise, such as coronary compression. The clinical course and various imaging modalities are useful for differentiating angiosarcomas from coronary stenosis.

## Keywords

Angiosarcoma • Mechanical complications • Ischaemic heart disease • Coronary spasm • Coronary intervention • Case report

## ESC Curriculum

3.1 Coronary artery disease • 6.8 Cardiac tumours • 3.4 Coronary angiography • 6.9 Cardiac dysfunction in oncology patients • 2.4 Cardiac computed tomography

## Learning points

- Patients with cardiac angiosarcoma can present with unstable angina, and progressive disease leads to many mechanical complications. Although extremely rare, compression of the tumour can lead to coronary artery stenosis.
- Intravascular ultrasound and computed tomography findings can be used to infer the cause of coronary artery stenosis with acute coronary syndrome.

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## Introduction

Angiosarcoma is one of the most common malignant cardiac tumours. This tumour proliferates rapidly and generally causes death through widespread infiltration of the myocardium, obstruction of flow within the heart, or distant metastasis.<sup>1–5</sup> Although complete surgical resection of these tumours is strongly desired, surgery is not usually an effective treatment because of the large mass of cardiac tissue involved or the presence of metastasis. Thus, patients with primary cardiac angiosarcoma who are inoperable undergo relatively less invasive treatments to improve their prognosis. In this study, we report a patient with cardiac angiosarcoma who underwent percutaneous coronary intervention (PCI) for severe coronary artery stenosis.

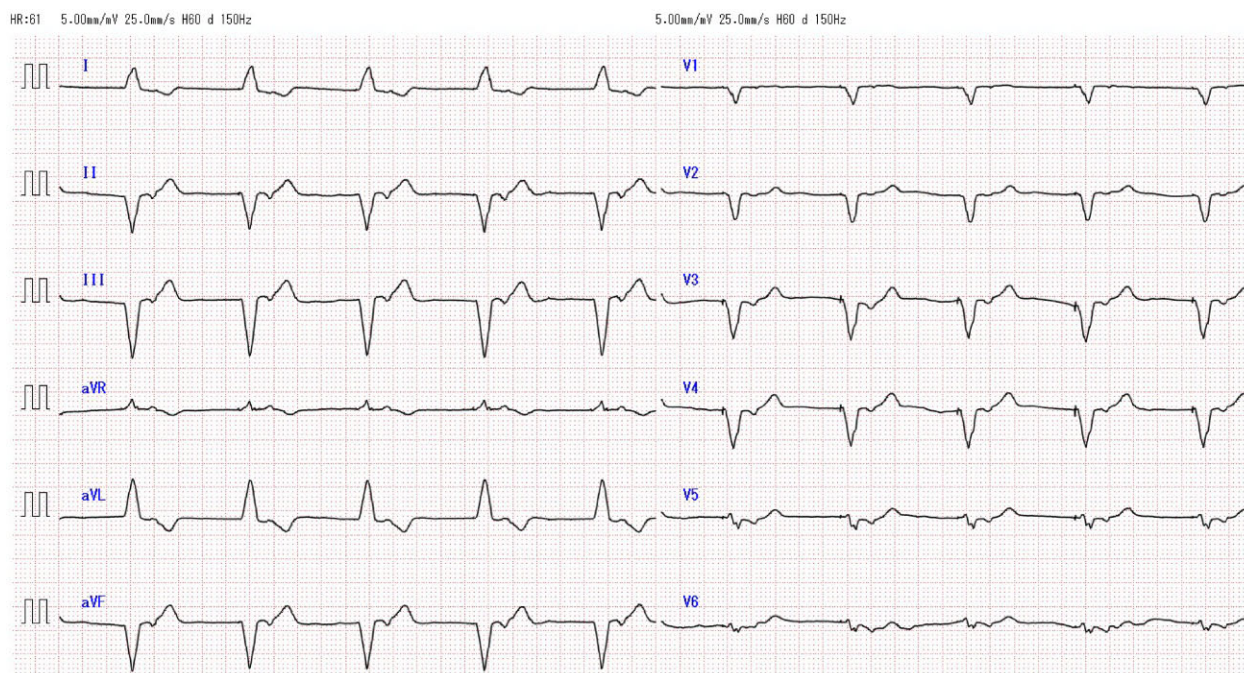
## Timeline

Date	Event
June 2018	Diagnosis of dyslipidaemia and hypertension. Presented with chest pain and heart palpitations. Patient diagnosed a right atrial angiosarcoma after having a pleuroscopic pericardial biopsy done.
July 2018	Patient underwent a lumpectomy to excise the tumour, but due to tissue adhesions in and around the right atrium, the malignancy could not be completely removed.
October 2018	Initiation of chemotherapy with Paclitaxel.
January 2020	Paclitaxel discontinued due to neuropathy. Started pazopanib.
June 2020	The patient was diagnosed with sick sinus syndrome and underwent implantation of a leadless pacemaker.
February 2021	Computed tomography (CT) showed tumour progression in the anterior mediastinum. Started eribulin.
17 March 2021	Emergency department presentation for progressively worsening chest pain. Emergency coronary angiography showed an isolated 99% ostial right coronary artery (RCA) stenosis. Intravascular ultrasound (IVUS) showed increased perivascular refraction at the tumour lesion was observed to push out the blood vessel from the short-axis direction. We performed drug-eluting stent deployment for prolonged severe stenosis of ostial RCA.
22 March 2021	Coronary CT scan showed that the tumour had progressed in the last several months at the site where coronary compression was observed by IVUS. All the investigations suggested a potential trigger of vascular stress and coronary vasospasm by tumour growth.
24 March 2021	Full recovery and discharge.
April 2021	Started trabectedin.
October 2021	Last follow-up. Patient in good clinical condition.

## Case presentation

A 64-year-old woman with chest pain and heart palpitations was referred to our hospital for further examination of a right atrial tumour. She had been prescribed oral medication for dyslipidaemia and hypertension. The tumour was diagnosed as an angiosarcoma, and surgical resection of the tumour was attempted. However, complete resection from the right atrium and area around the right coronary artery (RCA) could not be achieved because of its strong adhesion to surrounding myocardial tissue. Macroscopic residual disease was observed post-surgery, and adjunctive chemotherapy was started in the same year. Paclitaxel (100 mg/m<sup>2</sup>) and pazopanib (800 mg) were initially administered to the patient; however, computed tomography (CT) showed tumour progression in the anterior mediastinum. The patient was started on eribulin (1.4 mg/m<sup>2</sup>). During this period, the patient was diagnosed with sick sinus syndrome and underwent implantation of a leadless pacemaker. Three years after chemotherapy was started, the patient presented with chest pain and cold sweats at rest and was transported to our emergency clinic within an hour of onset. Although she complained of persistent chest discomfort, her vital signs were normal. Physical examination was unremarkable; there were no additional heart sounds on auscultation and no pulmonary rales. Jugular venous pressure was not elevated and no pitting oedema was present. Routine blood investigations on admission were unremarkable except for C-reactive protein 3.95 mg/dL (normal 0.0–0.14 mg/dL). Electrocardiogram showed worsening bradycardia (Figure 1), suggesting that the patient suffered from acute coronary syndrome. Emergency coronary angiography showed an isolated 99% ostial RCA stenosis with no other obstructive disease (Figure 2A). Intravascular ultrasound (IVUS) showed mild to moderate stenosis due to fibrous plaque formation with partial calcification of the vessels. We observed distortion of the blood vessel in the short-axis direction due to coronary artery compression at the tumour lesion (Figure 2B and C, Video 1). Additionally, the presence of extracardiac vascular blush was noted, supplied by right ventricular marginal branches. Initially, we suspected that the patient had coronary vasospastic angina, and repeatedly administered intracoronary nitroglycerine (10 mg) and isosorbide dinitrate (15 mg). Stenosis in the distal lesion was rapidly relieved by 2 mg of nitroglycerine, but treatment did not improve the ostial RCA stenosis (Figure 2D). Next, we successfully performed IVUS-guided PCI with drug-eluting stent deployment (Figure 2E and F, Videos 2 and 3). The Xience Skypoint stent (3.25 × 33 mm, Abbott Medical Inc., Abbott Park, IL, USA) was deployed from the ostium of the RCA to the distal lesion. A postoperative coronary CT scan showed that the tumour had progressed in the last several months at the site where coronary compression was observed by IVUS (Figure 3A and B).

The patient was taking aspirin 100 mg/day and clopidogrel 75 mg/day and was discharged from our hospital on postoperative day 7 without any further complications. The chemotherapy regimen was changed to trabectedin (1.0 mg/m<sup>2</sup>). Subsequently, her cardiac function was preserved, and no additional coronary events were observed after 7-month follow-up.



**Figure 1** Electrocardiography at the time of admission showed an all-pacing rhythm, suggesting ischaemic bradycardia.

## Discussion

Autopsy studies have suggested an incidence of primary cardiac tumours of 0.017–0.033%.<sup>3</sup> Most malignant cardiac tumours in adults are sarcomas, which often originate from the right atrium. Cardiac sarcomas account for 95% of malignant cardiac tumour cases. Primary cardiac angiosarcoma is the most common histologic subtype and constitutes 30% of these cases.<sup>4</sup> The mean survival time of this disease is  $3.8 \pm 2.5$  months without surgical resection<sup>5</sup> and there are no accepted treatment guidelines. Previous reports indicated a survival time of 12–30 months for patients who received various combinations of surgery, chemotherapy, radiation therapy, and/or transplantation.<sup>3,4</sup>

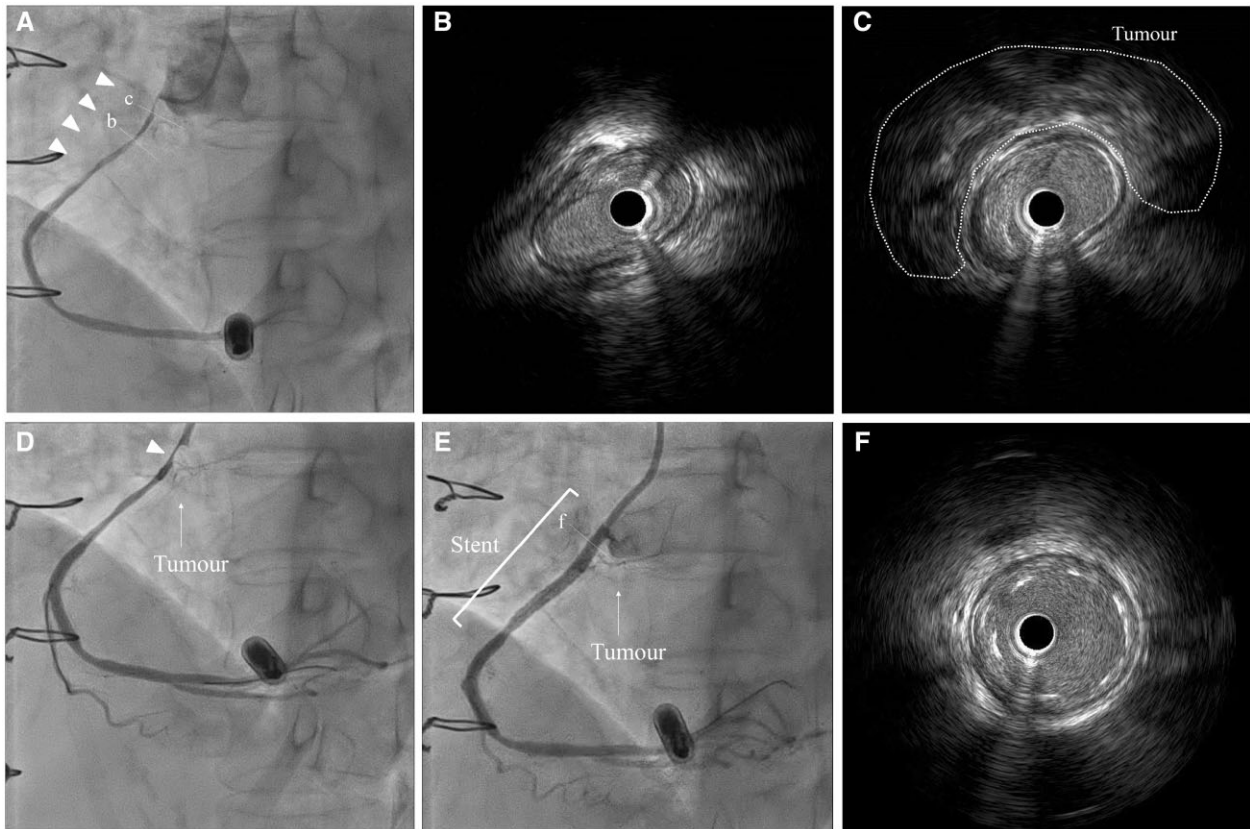
Many symptoms are non-specific and accompanied by various mechanical complications because cardiac tumours can arise in any part of the heart, and symptoms depend on the location and extent of the tumour. A retrospective analysis revealed distinct phenotypes in eight patients with primary sarcoma of the heart. Of those eight patients, two presented with pericardial effusion, two were initially suspected to have myxoma, and one experienced cerebellar infarction.<sup>1</sup> Coronary artery involvement is relatively rare, although tumours could interfere with valve function and blood flow through the heart. Most patients present with right-sided congestive heart failure or arrhythmias, pericardial effusion with cardiac tamponade, valvular dysfunction, tumour embolism, and diffuse pulmonary haemorrhage.<sup>2</sup>

Our patient was suspected to suffer from acute coronary syndrome and received catheter intervention, but it was initially difficult to diagnose coronary artery stenosis due to compression of the

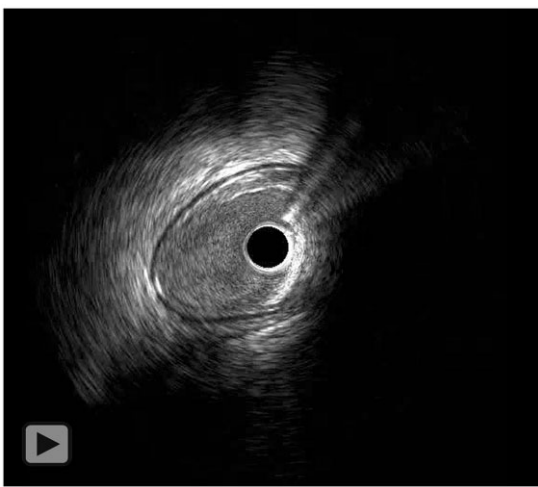
coronary artery by the cardiac tumour. Intravascular ultrasound showed calcification in areas containing fibrous plaques, but the overall vascular endothelial structure was preserved, thus the possibility of acute myocardial infarction with plaque rupture was excluded. At the beginning of treatment, we frequently administered nitroglycerine and isosorbide dinitrate for coronary vasospastic angina. Although it was relatively easy to reduce stenosis in the distal lesion, the ostial lesion was resistant to vasodilator agents. Intravascular ultrasound revealed perivascular compression from the tumour, and blood vessel deformation towards the short-axis indicated tumour progression.

Another possible mechanism of coronary artery involvement is coronary vasospasm due to a tumour. The tumour itself has been considered a potential trigger of vascular stress and coronary vasospasm, but there are no IVUS imaging findings of vasospastic lesions, such as diffuse intimal thickening,<sup>6</sup> negative remodelling,<sup>7</sup> or increased area of the sonolucent zone.<sup>8,9</sup> Thus, in this case, coronary artery compression was the major mechanism of the right coronary ostial lesion. A drug-eluting stent was deployed for the persistent severe stenosis of the RCA ostium.

Not many cases of acute coronary syndrome occur due to coronary artery compression by cardiac tumours.<sup>10–12</sup> In addition, various cardiac symptoms can be attributed to either direct coronary artery invasion and/or compression, tumour embolization, pericardial invasion and pericarditis, continued myocardial compression, or release of inflammatory mediators. Magnetic resonance imaging and CT could be useful for patients with a stable cardiovascular condition. If such imaging tests suggest coronary narrowing due to tumour



**Figure 2** (A) Emergency coronary angiography reveals 99% ostial right coronary artery stenosis with no other obstructive disease. (B) Intravascular ultrasound demonstrates mild to moderate stenosis due to fibrous plaque with partial calcification. (C) Distortion of the blood vessel in the short-axis direction due to coronary artery compression at the tumour lesion. (D) Stenosis in the distal lesion rapidly improved with 2 mg nitroglycerine treatment, but there was no effect on ostial right coronary artery stenosis. Extracardiac vascular blush was noted, supplied by right ventricular marginal branches. (E) Angio-images after stenting. An extracardiac vascular blush provided by right ventricular marginal branches was noted. (F) Intravascular ultrasound after treatment confirms successful expansion of the stent.



**Video 1** Pre-treatment intravascular ultrasound video.

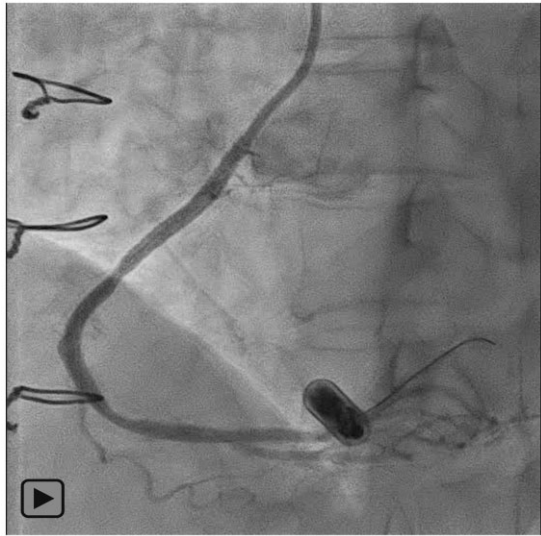


**Video 2** Post-treatment intravascular ultrasound video.

compression, the information would be helpful to determine the need for angiography or endovascular treatment.

## Conclusion

In conclusion, we present a rare case of angiosarcoma with coronary artery stenosis successfully treated with PCI. Coronary compression is one of the critical complications of advanced cardiac angiosarcoma.



**Video 3** Angiographic images after stenting.

## Lead author biography



Tadayuki Hirai is a cardiologist working at Ishikawa Prefectural Central Hospital, Japan. He is also an MD at the Graduate School of Medicine, University of Kanazawa, Japan. His main interests are structural heart disease and the role of sympathetic nerve activity in heart failure, especially in the context of intensive care and emergency medical care.

## Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

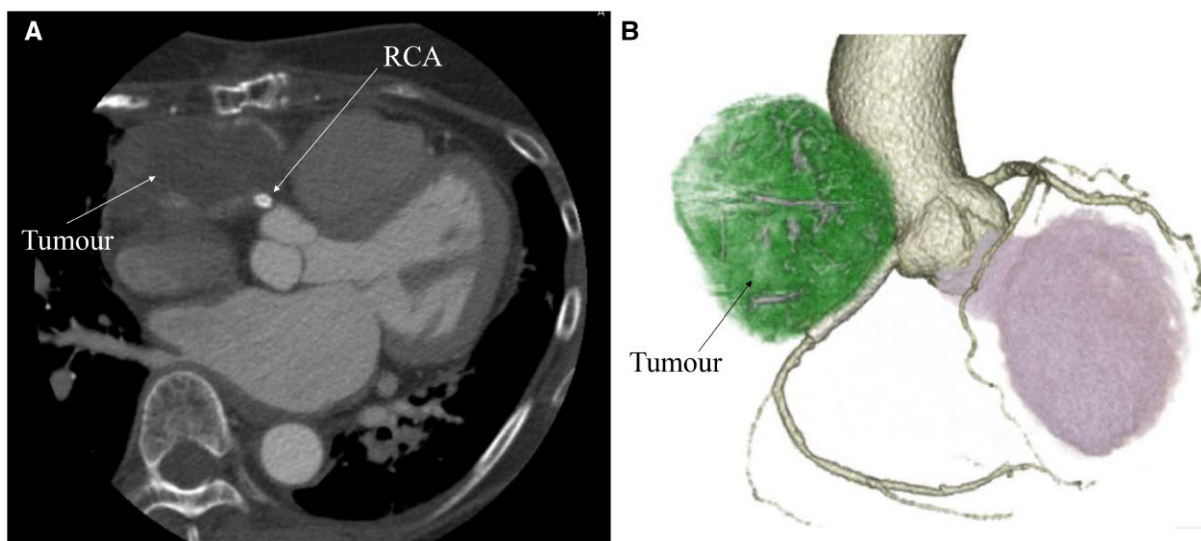
**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** None declared.

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**Figure 3** (A) Computed tomography scan shows tumour progression over several months at the site where coronary compression was observed by intravascular ultrasound. (B) Reconstructed coronary computed tomography images clearly depict the anatomical location of the tumour.

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