CASE REPORT

Successful treatment of primary cardiac lymphoma causing ST-elevation myocardial infarction by percutaneous coronary intervention combined with chemotherapy

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SUMMARY

A 76-year-old immunocompetent woman presented to our hospital with general fatigue. Her blood pressure was 60/40 mm Hg and pulse rate was 110 bpm. An electrocardiogram showed ST-elevation in the II, III and aVF leads with complete atrioventricular block. An echocardiogram and CT revealed pericardial effusion and a 6 cm solid tumour lying anterior to the heart. The right coronary artery (RCA) ran through the centre of the tumour, which bulged into the right atrium for 35 mm and vibrated. Emergent coronary angiography revealed 99% stenosis with delay at the proximal RCA; however, intravascular ultrasound showed no atheromatous changes, and the RCA was compressed by the extravascular mass. Successful coronary stenting improved the coronary flow. The following day, a biopsy was performed via thoracotomy without any events, the results of which showed diffuse-type large B-cell lymphoma histologically. Chemotherapy gradually reduced the tumour size, and the patient became stable haemodynamically.

BACKGROUND

Primary cardiac lymphoma (PCL) is a rare malignancy in immunocompetent patients. Cardiac complications are diverse and non-specific based on how and where the lymphoma involves the heart, and this variety in cardiac complications often leads to a delay in diagnosis. The incidence of PCL is currently increasing due to advancements in imaging modalities and the growing population of immunocompromised patients. Although there are several previous cases of PCL masquerading as acute coronary syndrome (ACS), in which ST-elevation and a high troponin-T level were due to myocardial infarction by the lymphoma, there are no reported cases of PCL causing 'real' ACS based on a disturbance of the coronary circulation. We describe the first reported case of ACS caused by PCL.

CASE PRESENTATION

A 76-year-old woman with no specific medical history presented to our hospital for faintness and general fatigue, with a blood pressure of 60/40 mm Hg, regular pulse rate of 110 per minute and oxygen saturation of 97% on air. The superficial lymph nodes were not swollen and there were no abnormalities, other than jugular vein distension, on a

physical examination. A chest X-ray showed cardiomegaly (cardiothoracic ratio, CTR, 58.1%). On an echocardiogram, there was significant pericardial effusion, and the right atrium was collapsed, which indicated cardiac tamponade. The echocardiogram also revealed a solid tumour, lying anterior to the right side of the heart, which bulged into the right atrium for 26×38 mm and were swinging (figure 1A1). Chest contrast-enhanced CT showed a 60×45 mm tumour with the right coronary artery (RCA) running through the centre of the lesion (figure 1B1). The patient was admitted to the intensive care unit and stabilised with fluid infusion. A biopsy via thoracotomy was planned for the next day; however, the patient fell into shock, and an electrocardiogram showed ST segment elevation in the II, III and aVF leads with complete atrioventricular block (AVB). Emergent coronary angiography (CAG) showed 99% stenosis with a thrombolysis in myocardial infarction (TIMI) 1 flow in the proximal segment of the RCA (figure 2A). Balloon angioplasty was performed using a perfusion balloon catheter with long inflation, which improved the coronary flow (figure 2A). At that time, stent implantation and the administration of antiplatelet agents were withheld because a biopsy via thoracotomy was planned for the following day. However, 3 h later, the patient's blood pressure decreased and her ST level increased. Re-CAG revealed acute occlusion of the RCA, while intravascular sonography demonstrated narrowing of the lumen due to oppression by the surrounding solid extracoronary tumour (figure 2C). Therefore, a bare metal stent was implanted in order to obtain a radial force against the compression induced by the tumour (figure 2B). The next day, the biopsy via thoracotomy was performed without any adverse effects. The pathological findings showed diffuse large B-cell lymphoma, and R-THP-COP therapy (rituximab, pirarubicin, cyclophosphamide, vincristine and prednisolone) was administered as chemotherapy to prevent cardiac toxicity, such as that caused by doxorubicin. This regimen gradually decreased the size of the tumour (figure 1A2, B2), and the patient became stable without any symptoms 15 months after the first admission. This case was previously reported in a case series, 'Therapeutic management in cardiac lymphoma', which focused on the management of lymphoma.¹ Therefore, we newly report this case, focusing on the importance of cardiac management.

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Figure 1 Echocardiography (A1) showed that the mass anterior to the heart was a solid tumour and the inside was heterogeneous. The mass bulged into the RA significantly, although it did not become trapped in the tricuspid valve. In the chest contrast-enhanced CT (B1), a large mass (60×45 mm) was detected anterior to the heart, bulging into the RA. The right coronary artery (arrow) ran through the centre of the mass. After 3 months, the mass anterior to the heart and the pericardial effusion almost disappeared and the size of the mass in the RA decreased (26×38→8×14 mm). Ao, aorta; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

DISCUSSION

Primary cardiac tumours are rare. Myxoma is the most common benign primary tumour, with sarcoma accounting for the majority of cases, while lymphoma is the second most common malignant lesion.^{2,3} The clinical manifestations of these tumours are diverse and non-specific based on how and where the lesion involves the heart. For example, myocardial infiltration may cause arrhythmia or myocardial damage, while a location in the epicardium may induce heart failure, cardiac tamponade or syncope and the involvement of the vascular system may stimulate the development of myocardial ischaemia and/or circulatory failure, including superior vena cava syndrome. Meanwhile, PCL accounts for approximately 1% of primary cardiac tumours and is much less frequent than secondary involvement. Moreover, it is rare for PCL to develop in immunocompetent patients, such as it did in the present case, accounting for only 1.3% of primary cardiac tumours.⁴ The survival of patients with PCL is affected by four factors: the immune status and the presence of left ventricular involvement, arrhythmia or extracardiac disease.⁵ AVB, which has been reported to be a common complication of PCL,⁶ was also detected in the present case. The patient's AVB improved following coronary artery revascularisation; therefore, the cause of AVB was

not PCL infiltration but, rather, myocardial ischaemia. There are two previous case reports of PCL mimicking acute myocardial infarction, in which ECG changes and troponin elevation were due to myocardial infiltration rather than infarction.^{7,8} In addition, a large case series (197 cases) was recently published in which there were no cases of PCL complicated with ACS.⁵ Therefore, the present article is the first report of PCL complicated with true acute myocardial infarction. Although most cases of acute myocardial infarction are caused by plaque rupture and/or the presence of thrombi in the coronary arteries, in the present case, intravascular ultrasound revealed no atheromatous plaque in the coronary arteries and demonstrated that the lumen narrowing was not caused by the thrombus, but was caused instead by the surrounding tumour mass, the expansion of which impaired the coronary flow, thereby inducing myocardial ischaemia (figure 2C). Immunoglobulin G4-related disease is also a common cause of lesions surrounding the coronary arteries;⁹ however, this abnormality was excluded based on histology in the present case. The prevalence of PCL is currently increasing due to advancements in imaging modalities and a growing population of immunocompromised patients. In addition, there are some reports of PCL in immunocompetent individuals, including the present patient.

Figure 2 On emergent coronary angiography (CAG), there was severe stenosis (arrow) in the proximal right coronary artery (RCA) with a thrombolysis in myocardial infarction (TIMI) 1 flow. Balloon angiography using a perfusion balloon improved the stenosis and the coronary flow (A). However, after 3 h, re-CAG showed acute recoil at the proximal RCA. On intravascular ultrasound (C), the stenotic lesion was oppressed by the surrounding mass and appeared to not be occupied by the thrombus. We implanted a stent (B, arrowhead).

Therefore, it is necessary to keep in mind the possibility of this lesion in cases involving cardiac or pericardial tumours.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Primary cardiac lymphoma (PCL) is a rare malignancy in immunocompetent patients.
- ▶ The cardiac complications of PCL vary according to how and where the lymphoma involves the heart, and this variety may cause a delay in diagnosis.
- ▶ This is the first reported case of acute coronary syndrome caused by PCL.
- ▶ It is difficult to perform coronary intervention followed by bleeding procedures, and this case highlights the challenges in treating similar cases in the real world.

Competing interests None.

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