Primary pericardial spindle cell sarcoma mimicking left main coronary artery disease

Joo Hyun Park, Hyunmin Choe*, Woo-Ik Jang and Gam Hur

a Department of Cardiology, Inje University College of Medicine, Ilsan Paik Hospital, Goyang, Korea
b Department of Thoracic and Cardiovascular Surgery, Inje University College of Medicine, Ilsan Paik Hospital, Goyang, Korea
c Department of Radiology, Inje University College of Medicine, Ilsan Paik Hospital, Goyang, Korea

* Corresponding author. 2240, Daehwa-dong, Ilsanseo-gu, Goyang-si, Gyeonggi-do 411-706, Korea. Tel: +82-31-910-7833; fax: +82-31-910-7829; e-mail: hmchoi49@naver.com (H. Choe).

Received 29 September 2011; accepted 4 November 2011

Abstract

Primary spindle cell sarcoma in the heart is a very uncommon disease. Although primary atrial or pulmonary vein spindle cell sarcomas have been sporadically reported, pericardial spindle cell sarcoma is rarely seen in currently available data. The commentary here is on a primary pericardial spindle cell sarcoma that was preliminarily misjudged to be left main coronary artery disease.

Keywords: Spindle cell sarcoma • Pericardium • Coronary artery disease

CASE REPORT

A 59-year old woman was admitted to our hospital with exertional chest pain and dyspnoea for 1 month. Her medical history included hypertension. Physical examination showed blood pressure at 140/80 mmHg, pulse rate at 54 beats/min and a respiration rate of 25 breaths/min. Heart sound and rhythm was regular. Initially, we diagnosed her with unstable angina and tested for coronary artery lesions by coronary angiography. Transcardial coronary angiography revealed a critical stenosis of ~90% in the left main coronary artery ostium (Fig. 1a). Right coronary artery angiogram showed collateral blood flow to the left coronary artery via the septal branch and minimal evidence of a coronary lesion at the ostium (Fig. 1b).

We diagnosed the patient as having definite left main coronary artery disease and deliberated on what corrective measure to choose, percutaneous coronary intervention or coronary artery bypass graft (CABG) surgery. We decided to pursue a second-stage percutaneous coronary intervention via the femoral route. However, routine echocardiography findings showed a moderate amount of pericardial effusion; this finding raised some doubts about an unusual coronary artery disease. Therefore, we performed coronary multidetector computed tomography (MDCT) angiography to evaluate the features of the coronary arteries and adjacent organs. Contrary to our first diagnosis of left main coronary artery disease, the multi-planar re-formation images of MDCT showed a 7.1 × 3.7 cm cardiac mass compressing the left main coronary artery ostium (Fig. 2a).

The cardiac mass was between the left atrium, aortic root and pulmonary artery and was well displayed and defined with heterogeneous enhancement. This mass was also encircling the left main coronary artery ostium (Fig. 2b).

The patient was treated by radical surgical excision of the tumour mass, and CABG was performed. An intraoperative macroscopic finding showed a lobulated and firm intrapericardial mass measuring 11.5 × 7.2 × 5.4 cm (Fig 2c). The mass originated from the posterior wall of the aorta and pulmonary artery portion and extended leftwards to the atrial auricle and superior vena cava. Particularly, the infiltrating mass was encircling and compressing the left main coronary artery ostium. We dissected and removed the tumour mass that was invading and extending around the aorta, left atrium, pulmonary artery and left main coronary artery, as completely as we could. Subsequently, following this, anastomoses of the left internal mammary artery to the left anterior descending artery and the saphenous vein graft to the left circumflex were performed. Microscopic examination revealed that the tumour was composed of spindle cells exhibiting non-specific differentiation (Fig. 2d). These findings led us to the diagnosis of primary cardiac spindle cell sarcoma. Also, mediastinal and hilar lymph node biopsy confirmed tumour cell infiltration. The patient is alive 12 months after surgery while continuing with both radiation therapy and chemotherapy, with no sign of local recurrence and metastasis.

DISCUSSION

Similar to metastatic cardiac tumours, primary cardiac spindle cell sarcoma is a very rare tumour [1]. A primary cardiac spindle cell sarcoma can occur in the great vessels, pulmonary veins, the right and left atrium [2], but its occurrence in the pericardium has only been reported in one case report [3]. This tumour can have clinical manifestations by obstructing forward flow in the great vessels or cardiac valves, and causing arrhythmias, embolic
events and distant metastases [4]. In our case, we would have misdiagnosed it as left main coronary artery disease and unwittingly carried out percutaneous coronary intervention due to typical anginal pain and coronary angiographic findings.

Radical surgical resection was the optimal choice for prolonged survival compared with those with incomplete surgical clearance [5]. Because this present case has shown extensive invasion by the tumour with infiltration of both surrounding tissues and mediastinal lymph nodes, we justly added adjuvant chemotherapy and radiation therapy. However, we expect the prognosis of such patients to be very poor because the tumour can be resistant to chemotherapy and radiotherapy [2]. Actually, there is still evidence of local recurrences and metastasis in spite of adjuvant chemotherapy and radiation therapy for this patient.

The most common cause of death, notwithstanding complete surgical removal, is local recurrence of the primary tumour in ≏50% of patients. Also, with any of the different treatment modalities that we may choose, the prognosis is ominous after diagnosis [6]. However, precise rates for survival, metastasis and recurrence specific to primary pericardial spindle cell sarcomas are not available at this time.

CONCLUSION

We have described a case in which we incorrectly considered a diagnosis of left main coronary artery disease due to typical chest pain symptoms and coronary angiography findings. Only
after further scrutiny and MDCT angiography did we reach the correct diagnosis: primary cardiac spindle cell sarcoma.

Conflict of interest: none declared.

REFERENCES


