Leiomyosarcoma with coronary fistulae and ventricular septal perforation: A case study

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Abstract

Coronary fistulae and ventricular septal perforation are very rare clinically, and even less caused by cardiac leiomyosarcoma. A case is reported that a 67-year-old female had cardiac leiomyosarcoma with progressive heart failure and coronary fistulae and ventricular septal perforation. This case was special since all ante-mortem examinations and cardiac surgery failed to detect the presence of any abnormal cardiac mass. Therefore, the malignant cardiac tumors could appear in an invasive form without mass and be one of the causes of the coronary fistulae and ventricular septal perforation.

Keywords: Cardiac leiomyosarcoma; Coronary fistulae; Ventricular septal perforation; Heart failure

1 Introduction

Primary cardiac malignancy is very unusual with an estimated incidence ranging from 0.001% to 0.03%[1] nine percent of which, are ascribed to cardiac leiomyosarcoma (CL), making it extremely rare. CL often has physical mass. Diagnoses are most often made after a mass in the chambers of a heart is found or abnormal cardiac hemodynamic caused by an occupying tumor is noticed. While the exact oncogenesis is not known, CL is highly progressive and locally invasive.[2] It can be either a primary tumor in heart or a metastatic tumor arising from other primary sites.[3,4]

2 Case report

A 67-year-old woman was admitted with recurrent chest tightness for four years, progressive dyspnea for six months, and nausea for two months. The past history included hypertension for over 30 years, and diabetes and hyperlipidemia for two years. A left mastectomy for breast cancer was performed 19 years ago, followed by local radiation therapy of three months. There was no evidence of recurrent disease.

Her physical examination revealed the blood pressure of 105/80 mmHg and the heart rate of 118 beat/min in sinus rhythm. Moist rales were audible in both lungs. The heart edge enlarged towards the left. A grade IV pan-systolic murmur could be heard at the lower left sternal edge.

Complete right bundle branch block was shown on her electrocardiogram. Her chest X-ray showed significant cardiomegaly with interstitial edemas in both lungs. Echocardiography (Figure 1) showed the hypokinesis in the middle to apex sections of the left ventricle and the paradoxical septal motion in the apex while the continuous interruption in the septum between the apex and middle sections with left to right shunting was observed, and left ventricular ejection fraction (LVEF) was 34%. The echocardiography performed three years ago demonstrated mild hypertrophy of the ventricular septum, normal myocardium motion and heart chambers, and LVEF from 58% to 65% while the recent echocardiographic results indicated the hypokinetic development of her left ventricular walls and LVEF from 43% to 48%. A coronary artery angiogram showed atherosclerosis in the mid-left anterior descending artery (LAD), and the luminal stenosis of 60% compared with 50% three years ago. There were multiple coronary fistulae in the LAD draining into the left ventricle and fistulae in the end of left circumflex (LCX) and the right coronary artery (RCA), compared with small left coronary fistulae in the LAD and none in the LCX and RCA three years ago. The angiogram
also displayed the left ventricular septal perforation. The single photon emission computed tomography (SPECT) carried out two years ago illustrated an irregular radionuclide distribution in the anterior, inferior, posterior and septum segments of the myocardium with no evidence of ischemia after exercise.

The patient was initially diagnosed and treated in accordance with coronary artery disease. However, it was unsuccessful. Her symptoms worsened and progressive heart failure developed. Cardiac resynchronization therapy was performed to improve her progressive congestive heart failure, but the beneficial effect was not sustained. An intra-aortic balloon pump (IABP) was inserted, which improved her conditions significantly. However, she proceeded to worsen after the removal of IABP. The IABP was inserted again and the patient underwent surgical repair of her ventricular septal defect.

At the operation, her ventricular myocardium appeared dark grey and ventricular walls were thinner. A left apical aneurysm was present, but no mass was found in the chambers. A ventricular septal defect measuring 10 mm in diameter near the apex was identified. The aneurysm was removed and the defect was patched. Myocardial biopsy was taken from her ventricle myocardium and sent for histopathological analysis. The myocardium motion was not recovered and the patient died from refractory cardiac failure one week post operatively.

The histopathology report of the myocardial specimen confirmed the presence of malignant spindle cell neoplasm with prominent pleomorphism (Figures 2). There were signs of mitotic activities with atypical features with coarse grain chromatin, thin cytoplasm and diffuse foliated necrosis. Immunohistochemical stains were positive for vimentin, smooth muscle actin, S-100 and caldesmon, and negative for pan-cytokeratin, myoglobin, CD34 and CD 99. These findings were consistent with the diagnosis of the CL.

### 3 Discussion

The patient presented the progressive development of
chest tightness and heart failure, and did not positively respond to the treatment in accordance with myocardial ischemia. The myocardial hypokinesis and the heart function impairment were observed in the echocardiography. SPECT demonstrated the irregular radionuclide distribution in the myocardium segments, which was not bonded to the myocardial ischemia. The above observations illustrated that the abnormality of the myocardium existed and may be related to the invasion of the CL. The infiltration of malignant cells into the myocardium could cause the injury of the myocardium, resulting in the abnormal motions of the ventricular walls and the ventricular septal defect. Furthermore, progressive coronary fistulae also resulted from the invasion of the tumor. The coronary fistulae resulted in the ischemia at the end of the myocardium, which enhanced the abnormal motions, and further led to the formation of the ventricular aneurysm and ventricular septal perforation.

The uniqueness of this case is that various examinations and even the surgical investigation failed to recognize the presence of cardiac mass or neoplasm. The final diagnosis of the CL was made from the myocardial biopsy and the CL originated most likely from the heart. The results of the Positron Emission Tomography (PET) and tumor marker examinations were normal, which led to no evidence of the CL from other tumor metastasis. To date, there has not been any report of a cancer caused by local breast cancer radiotherapy. The radiation destruction or radiotherapy has not been known to cause etiology of the CL. Thus, it is impossible that the CL was caused by the local breast cancer radiotherapy.

To our knowledge, this is the first reported case of CL associated with multiple coronary fistulae and ventricular perforation, but without a mass.

In conclusion, this case is the primary CL characterized by the progressive congestive heart failure and coronary fistulae and ventricular septal perforation without a mass, which has not been previously reported. The lesson learned was that the possible existence of a myocardial disease or even CL, should be suspected if the presentation of usual coronary heart disease with congestive heart failure but with poor response to the corresponding conventional therapy has existed.

**References**

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