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Hidden left ventricular sarcoma manifesting itself with cardiogenic shock

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Abstract

Cardiac sarcomas are rare and aggressive tumors that could have a multiorgan involvement and unfavorable prognosis. We present an extremely rare situation of cardiac sarcoma in a fragile elderly patient with a dramatic presentation of cardiogenic shock.

KEYWORDS

cardiac sarcoma, cardiogenic shock

1 | INTRODUCTION

Primary cardiac sarcomas are rare and aggressive tumors with an unfavorable prognosis. They remain asymptomatic until they become locally advanced or until metastasis occurs. They produce nonspecific symptoms such as chest pain, dyspnea, and congestive heart failure.[1] Management of cardiac sarcomas includes complete surgical resection, when feasible, while the role of adjuvant and neoadjuvant therapy is not well defined so far.[1,2]

2 | CASE REPORT

This is the case of an 81-year-old woman who arrived at the emergency department presenting cardiogenic shock and pulmonary edema. Her blood pressure was initially 90/60 mmHg. The first arteriosus hemogasanalysis showed severe metabolic acidosis (pH 7.22; lactic acid 9 mMol/L). ECG showed sinus tachycardia at 120 bpm, low voltages in precordial leads, and diffuse nonspecific repolarization abnormalities. The X-ray chest revealed cardiomegaly and pulmonary congestion (Figure 1A). The patient was immediately transferred to the Cardiological Intensive Care Unit.

Transthoracic echocardiography showed a moderate left ventricular dysfunction (FEbp 38%) with akinesia of the inferior-lateral wall and hypokinesia of the remaining segments; right ventricular global function was also depressed (RV-FAC 30%). There was also severe mitral regurgitation and moderate tricuspid regurgitation with an estimated moderate pulmonary hypertension (sPAP 45 mmHg). Hidden in the inferior-lateral left ventricular wall, a mass of 9 × 6.5 cm was present, tangled between the left ventricular lateral wall and the pericardium (Figure 1A,B).

Despite pH correction with bicarbonate and the use of increasing doses of inotropes, the patient became anuric and rapidly developed multiorgan failure. The case was discussed by the Heart Team. In consideration of the fragility of the patient, difficulties achieving hemodynamic stabilization, and the poor short-term prognosis, surgery and/or other invasive examinations, such as coronary angiography, were decided against.

In less than 24 h, the patient became progressively bradycardic and entered cardiac arrest. Despite prolonged cardiopulmonary resuscitation maneuvers, the patient did not recover.

The day after, an autopsy defined the leading cause of death. The cardiac pathologist found a huge cauliflower mass (Figure 2) that

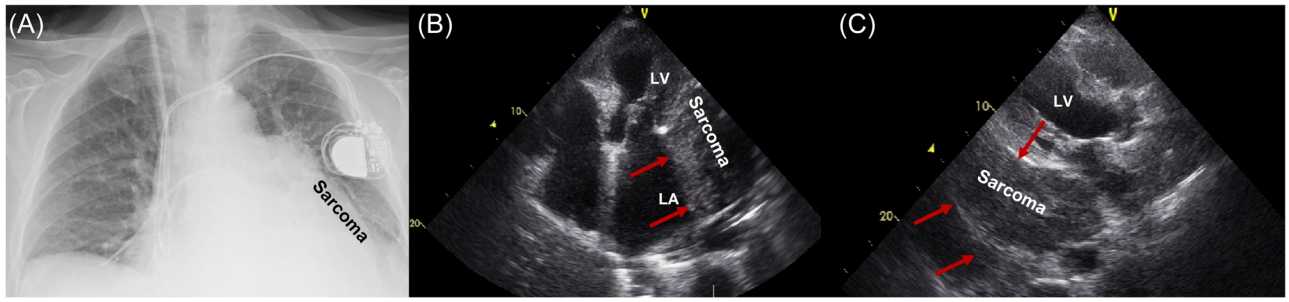

FIGURE 1 Chest X-ray (A) and transthoracic echocardiography (B and C) shows a huge cardiac sarcoma surrounding the left ventricle at the inferior-lateral wall.

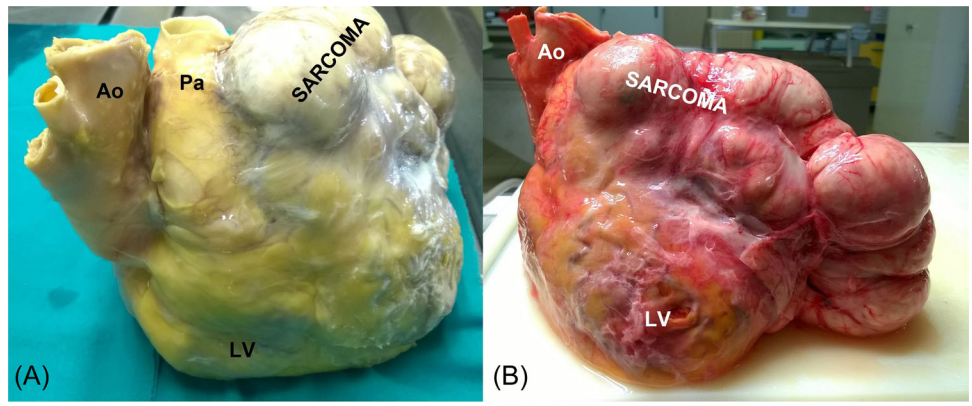

FIGURE 2 The gigantic cauliflower sarcoma with a polylobate shape extended from the left atrium to the proximity of the pulmonary artery. Ao, aorta; LV, left ventricle; Pa, pulmonary artery

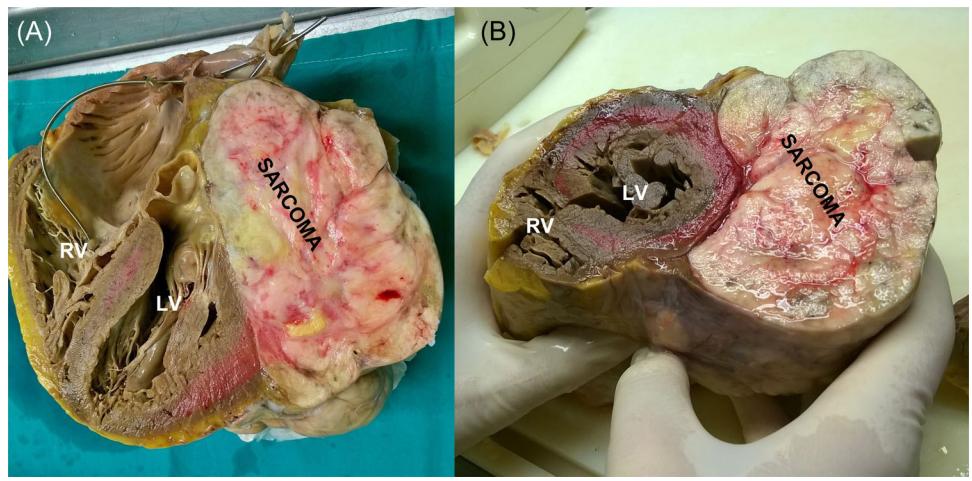

FIGURE 3 Cut sections are (A) long axis four-chamber section and (B) short-axis section at the mid-basal level (LV, left ventricle; RV, right ventricle). The sarcoma surrounded the left ventricle at the inferior-lateral wall but did not infiltrate the left ventricle, separated from it by a connective capsule.

surrounded the left ventricle (Figure 3). The heart weighed 1530 g and had a transverse diameter of 18 cm while a normal weight of an adult human heart is 300 g with a transverse diameter of 10 cm. The tumor had a polylobate shape and it was extended from the left atrium to the proximity of the pulmonary artery. The tumor did not infiltrate the left ventricle but rested on the ventricle, separated by a connective capsule. The brown color of the left ventricle was secondary to formalin fixation while the red areas of the left ventricle were areas of inappropriate fixation. In fact, formalin was injected inside and outside the heart cavities. The tumor showed large

polycentric-polyfocal necrotic zones. The red and yellow areas represented areas of tissue necrosis. The vital portion of the tumor was light gray, and the connective tissue capsule had a transparent greyish color.

Tumor compressed the circumflex artery and its collaterals (posterolateral and marginal ones; Figure 4). Acute ischemic distress was histologically visible in the left ventricle at the inferolateral level. The tumor, instead, did not have an ischemic process.

Moreover, there was moderate muscular hyperplasia of the pulmonary arterial vessels as per pulmonary hypertension and sub-centimetric pulmonary micrometastases were found.

Biopsy specimens (Figure 5) revealed a desmin and alpha-SMA positivity, consistent with a poorly differentiated pleomorphic leiomyosarcoma.

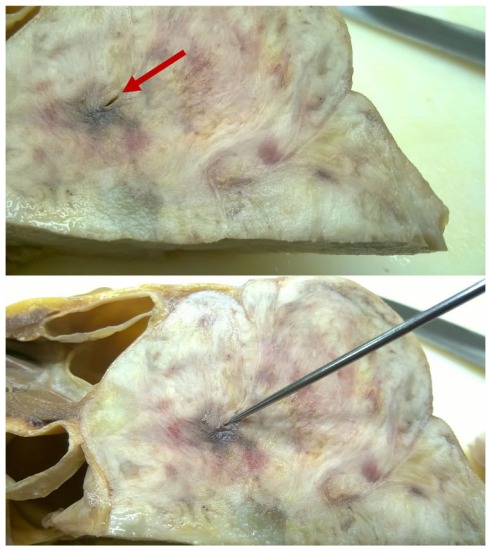

FIGURE 4 Red arrow shows the compression on the circumflex coronary artery.

3 | DISCUSSION

Primary cardiac tumors are extremely rare. They have an incidence of 0.001%–0.003% in the autopsy series. Nearly 25% of primary cardiac tumors are malignant and sarcomas are the most common histology. Almost half of all cases are vessel-derived tumors (e.g., angiosarcomas).[2,3]

Cardiac sarcomas have a poorer prognosis than noncardiac sarcomas. In fact, the median overall survival for patients with cardiac sarcomas is 6 months instead of 93 months for patients with noncardiac sarcomas.[2]

Surgery represents the first line of treatment for cardiac sarcomas. Patients who undergo surgery have a better prognosis (median survival of 12 months) while patients who do not undergo surgery have a shorter median survival (1 month). Surgery can also be performed for palliative debulking.[2] In our case, the surgical removal

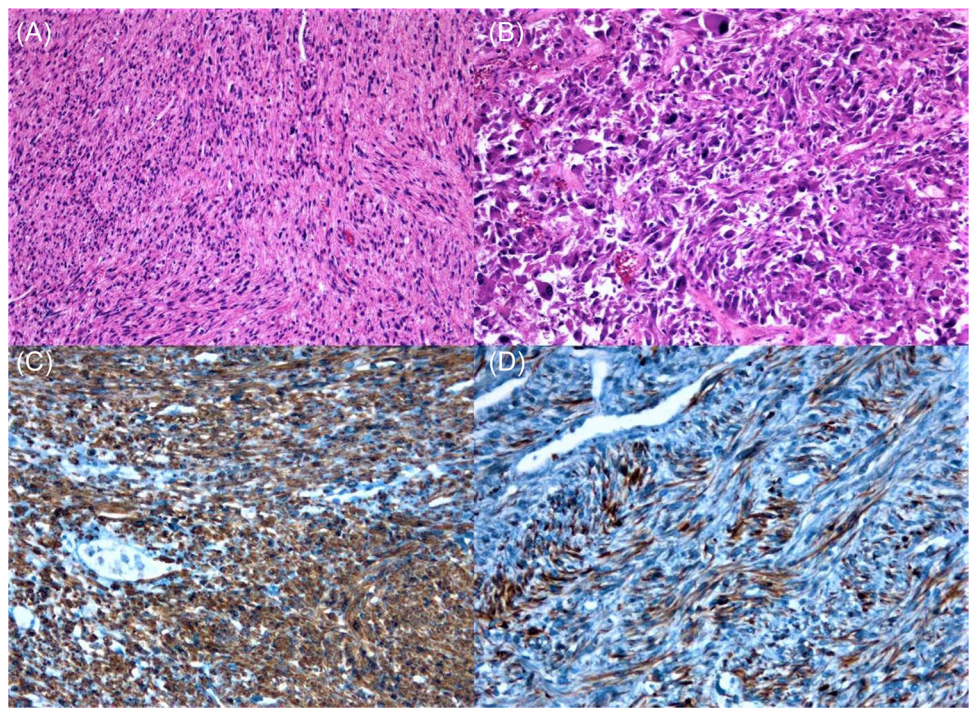

FIGURE 5 (A) Haematoxylin and eosin stain at ×10 magnification. Malignant mesenchymal spindle cell proliferation with cigar-shaped nuclei. (B) Biopsy specimen stained with hematoxylin and eosin. In some areas, the neoplasm has pleomorphic and bizarre cellular features. ×10. (C and D) Since the tumor was intensely positive for smooth muscle actin and focally positive for desmin, consistent with a poorly differentiated pleomorphic leiomyosarcoma ×20.

of the mass was not performed due to the severe condition of our patient and the difficulty of achieving good hemodynamic status. However, if a good hemodynamic status is obtained with mechanical and pharmacological support, surgical removal can take place even in urgent cases.[4,5] Our case report represents an extremely rare situation, both for the dramatic presentation of cardiogenic shock and for the atypical age of manifestation. In fact, cardiac sarcomas are more frequently present in the fourth and fifth decades of life.[2]

4 | CONCLUSION

Cardiac sarcomas are rare entities. While nonspecific symptoms are sometimes associated with sarcomas, the first presentation with cardiogenic shock is even rarer.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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