

CASE REPORT

Pericardial Synovial Sarcoma presenting with Cardiac Tamponade

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ABSTRACT

Pericardial synovial sarcomas are very rare and usually associated with poor prognosis if not diagnosed and treated early. It usually presents with symptoms of a massive pericardial effusion and signs of cardiac tamponade and failure. Here, we report the case of a 27-year-old patient with exertional dyspnea and massive pericardial effusion. Echocardiography and magnetic resonance imaging revealed a pericardial mass. The patient was treated by surgical excision and a subsequent histological analysis confirmed the diagnosis of pericardial synovial sarcoma.

Keywords: Cardiac tamponade, Pericardial effusion, Pericardial synovial sarcoma, Pericardium.

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INTRODUCTION

Synovial sarcoma of the pericardium is a rare entity.^{1,2} It is clinically rare and has various different histopathological appearances. Diagnosing this rare disease may be difficult. It is often confused with various other pericardial diseases. The patient may show a pericardial tumor with effusion on ultrasonography and chest computerized tomography (CT). It may present with symptoms and signs of cardiac tamponade. Diagnostic imaging may reveal a pericardial mass. Biopsy may be required to diagnose this condition.^{2,3} Molecular biological studies may help in the diagnosis and show reciprocal chromosomal translocation (X; 18 p11.2; q11.2).^{3,4} We report the case of a 27-year-old patient with exertional dyspnea who was operated and finally diagnosed with primary pericardial synovial sarcoma by histopathology and also by immunohistochemistry.

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CASE REPORT

A 27-year-old male presented with breathlessness on exertion. On examination, heart sounds were muffled. The chest X-ray indicated cardiomegaly (Fig. 1). Two-dimensional (2D) echo showed multiple intracardiac tumors – in left atrium (LA) and right atrium (RA), attached to septal free wall, and to mitral leaflets (Fig. 2). Tumor was seen extending outside the heart. Surrounding the heart and the tumor, a huge pericardial effusion was also present (Fig. 3). The CT scan confirmed the findings (Fig. 4).

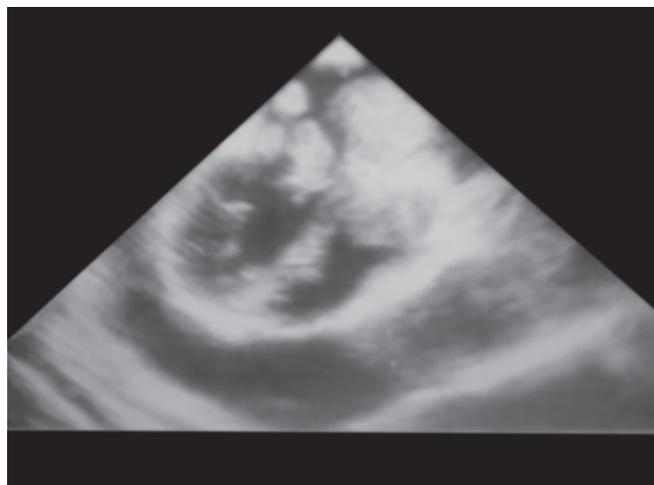


Fig. 1: X-ray chest showing cardiomegaly



Fig. 2: Contrast-enhanced CT images, showing localized pericardial effusion with septation and the thickened pericardium

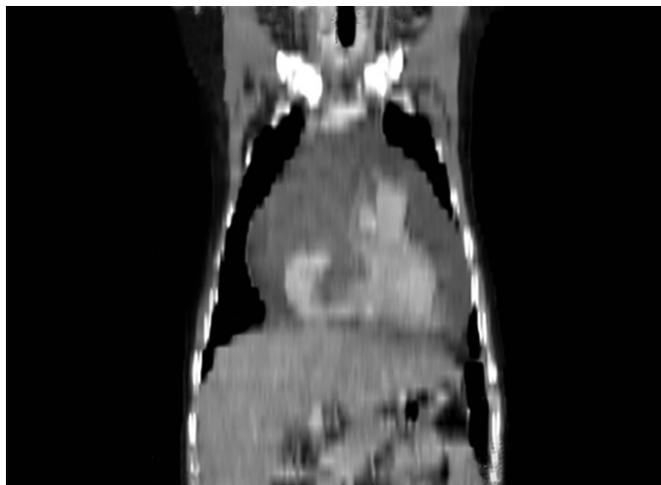


Fig. 3: Parasternal long axis view showing pericardial effusion

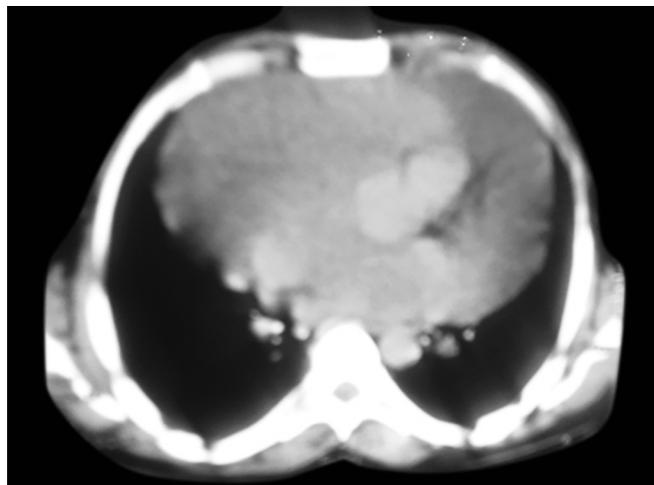
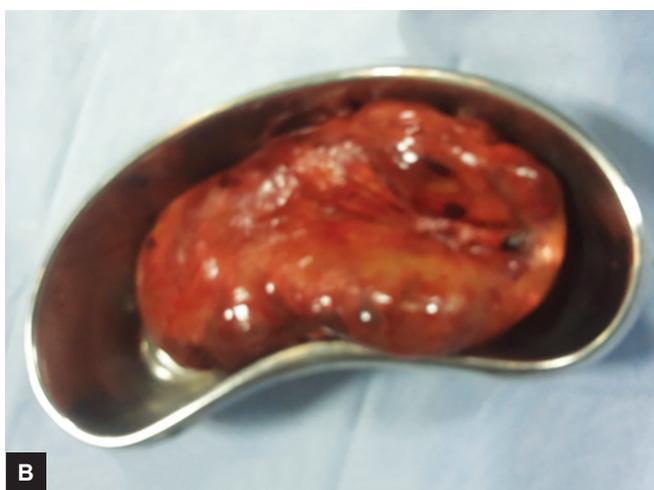
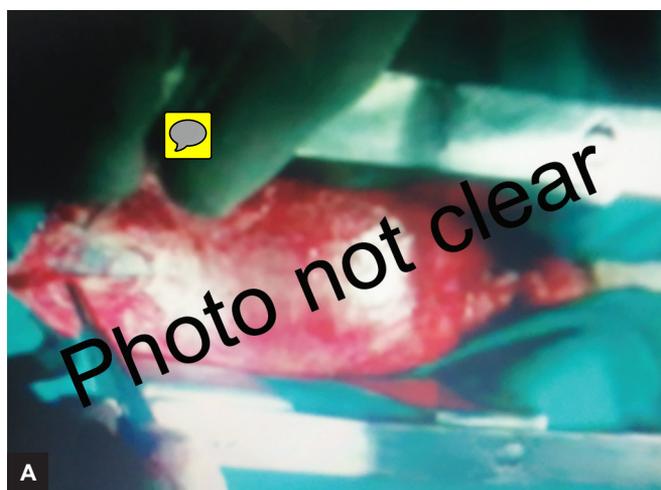


Fig. 4: Cardiac chest axial CT angiographic findings



Figs 5A and B: The tumor is located in the pericardium. Gross specimen shows a large, well-demarcated tumor

Soon, the patient developed severe breathlessness and was intubated. The 2D echo showed increase in pericardial effusion causing tamponade.

Patient was taken for open heart surgery. On sternotomy, heart was found to be entirely covered with tumor and none of the cardiac chambers were visible.

Femoral arteries were exposed to go on cardiopulmonary bypass (CPB). Tumor was gently cleared off in front of aorta. Tumor was found to be firm in consistency, but friable. Aortic cannulation was done after heparinization. Tumor was cleared gently from superior vena cava, inferior vena cava, and RA. Selective venous cannulations were done and snared. Rest of the cardia was cleared of tumor.

The CPB was established. The RA and LA were opened and multiple tumor growths were removed (Figs 5A, B and 6). After thorough wash, chambers were closed. Patient was weaned off CPB. He was electively ventilated for 1 day and then extubated.

Histopathology showed synovial sarcoma of heart. Under high-power examination, a characteristic biphasic

appearance consisting of hypercellular spindled cell sheets was observed, which was confirmed by immunohistochemistry. Analyses by immunohistochemistry showed positive B-cell lymphoma (bcl)-2 and CD99.

Patient was referred to oncology department for further evaluation and necessary chemotherapy and radiotherapy (Fig. 6).

DISCUSSION

In this report, we have described the case of a patient with a pericardial synovial sarcoma, which is an extremely rare condition.¹⁻⁴

A 27-year-old male presented with breathlessness on exertion. On examination, heart sounds were muffled. The chest X-ray indicated cardiomegaly. The 2D echo showed multiple intracardiac tumors – in LA and RA – attached to septal free wall, and to mitral leaflets. Tumor was seen extending outside the heart. Surrounding the heart and the tumor, a huge pericardial effusion was also present.



Fig. 6: Patient before discharge

The 2D-echocardiography showed a large pericardial effusion. The chest CT revealed a pericardial cystic mass, effusion with pericardial thickening.

The most common benign pericardial tumors are pericardial cysts and lipomas. Mesothelioma is the common primary malignant tumor of the pericardium. Other malignant tumors include a large variety of sarcomas and lymphomas. Cheng et al⁴ have described histologic subtypes of pericardial sarcoma as angiosarcoma, undifferentiated sarcoma, fibrosarcoma, liposarcoma, rhabdomyosarcoma, and synovial sarcomas.⁵

Pericardial synovial sarcoma is very rare and only 20 cases of synovial sarcoma of the pericardium have been reported in the literature¹⁻³ so far.

Histologically, synovial sarcoma has three subtypes: biphasic, monophasic, and poorly differentiated types. The tumor has a slight predilection for the male sex; the average age at presentation is around 30 to 35 years. The common symptoms are dyspnea on exertion and signs of cardiac tamponade due to pericardial effusion.^{1,3} The quantity of pericardial effusion is variable and most cases show a pericardial mass, while fewer have no visible mass.³

The X-ray of the chest is the first tool of choice for the differential diagnosis of dyspnea. Echocardiography or chest CT will help us further in confirming the diagnosis.⁸ It has to be distinguished from other types of sarcomata, which are malignant tumors.^{1,6,7} A cytogenetic analysis after surgical biopsy confirmed the diagnosis of pericardial synovial sarcoma in our case. The immunological finding of bcl-2 and CD99+ may be seen in other tumors also.³

The presence of the fusion gene *SS18-SSX* by the reciprocal translocation $t(X; 18)(p11; q11)$ is a hallmark cytogenetic finding.

As the tumor is rare and highly aggressive in behavior, treatment strategies have not been identified yet. These

tumors result in a median survival of 2 to 3 years.^{1,2} In the reported cases, complete excision or near total removal was possible in only 17 to 22.7% of cases. Chemotherapy and radiation therapy are recommended as adjuvant therapy for residual or recurrent tumors.^{1-3,9,10}

Sagrìstà-Sauleda et al¹¹ have suggested that presence of hemorrhagic pericardial effusion or cardiac tamponade is the diagnostic clue for pericardial malignancy.

CONCLUSION

To conclude, whenever idiopathic pericarditis is suspected because of a large amount of nonhemorrhagic pericardial effusion, a rare pericardial malignancy should be considered in the differential diagnosis.

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