Complete resection of giant pericardial synovial sarcoma in a 7-year-old boy: a case report

Qingyun Liu[1], Zhenhua Huang[1], and Sihai Gao[1]

[1]Tongji Hospital of Tongji Medical College of Huazhong University of Science and Technology

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Abstract

Background: Synovial sarcoma is a rare soft-tissue malignant tumor most commonly occurring in the extremities and head and neck region, and rarely occurring in the pericardium. Case presentation: We report a 7-year-old boy was admitted to the hospital with recurrent fever and chest pain over the past four months. A cardiac magnetic resonance imaging (MRI) revealed a tumor beneath the heart in the pericardial, and we surgical resection it completely. The postoperative histopathological examination resulted in a diagnosis of monophasic spindle cell type synovial sarcoma. After two weeks of hospitalization, the patient was discharged. Three months after discharge, the positron emission tomography (PET/CT) scans did not show any signs of recurrence. Conclusion: Pericardial synovial sarcoma is a rare disease that is detected early, and complete resection improves patient survival. We recommend CT be performed in patients with recurrent fever and sizeable pericardial effusion to rule out possible pericardial synovial sarcoma considering the echocardiography limitations.

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Qingyun Liu[1]; Zhenhua Huang[1]; Sihai Gao[1]

[1]Divison of Cardiothoracic and Vascular Surgery, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, China.

Correspondence:

Sihai Gao, Divison of Cardiothoracic and Vascular Surgery, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, China.

Email: *sihaigao73@163.com*

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Keywords

Pericardial; synovial sarcoma; heart neoplasms; case report

Background

Synovial sarcoma is a rare soft-tissue malignant tumor. Pericardial synovial sarcoma (PSS) is a sporadic disease with a dismal prognosis (1). Most patients present with dyspnea, cough, and chest pain caused by heart failure (2). According to JOSE Duran-Moreno's analysis from 37 cases reported in English during the past 20 years, PSS was found to occur in the 13-67 age range (3). To our knowledge, our case is younger than previously reported. In this letter, we report a case of the enormous pericardial synovial sarcoma in a 7-year-old boy.

Case presentation

A 7-year-old boy was weighing 22 kilograms, with no meaningful medical history, presented with fever and chest pain four months ago. A small amount of pericardial effusion was found in the local hospital, and the symptoms were relieved after treatment. Two months later, however, these symptoms reappeared. They went to another hospital, where their symptoms were reduced after treatment. After echocardiography and Computed Tomography (CT) revealed a large mass in the pericardial cavity the patient was referred to our institution. The patient had no additional significant medical, surgical, or family history. An echocardiogram revealed a large pericardial mass with pericardial effusion and right heart enlargement. Fortunately, the ejection fraction was in the normal range of 69%. Doppler echocardiography revealed a huge irregular hypoechogenic area seen in the pericardial cavity, ranging from about 144 × 95mm, and irregular anechoic area could be seen(Figure 1.A). A cardiac Magnetic Resonance Imaging (MRI) demonstrated a 142x94x81-mm mass below the heart in the pericardial cavity and close to left and right ventricles and inferior wall of left and right atria (Figure 1.B), partial enclosure of inferior vena cava (IVC).

We perform surgical resection of the mass by median sternotomy. The histopathology showed a monophasic tumor of spindle cells (Figure 2). Immunohistochemistry: EMA (partial +), VIM (+), tle-1 (+), INI1 (+), PCK (-), CK8 / 18 (-), CD117 (c-kit9.7) (-), CD117 (positive control) (+), CD34 (vascular +), dog1 (-), SMA (-), DES (-), MyoD1 (-), myogenin (-), caldesmon (-), S-100 (-), Sox10 (-), SDHB (-), STAT6 (-), Ki-67 (hot spot Li˜ 40%). Molecular testing: ss18 FISH (+); ETV6 (-). Two weeks from the resection, the patient was discharged and declined adjuvant therapy in our hospital. Three months later, PET/CT has not shown any evidence of recurrence or metastasis.

Discussion and conclusions

Primary cardiac and pericardial tumors are extremely rare entities at an incidence of 0.001% to 0.03% in autopsy series, and most of them are benign tumors (4, 5). From our center's retrospective data, malignant tumors accounted for about 10% of cardiac and pericardial tumors in the past ten years. Primary pericardial tumors are even less common, accounting for only 6.7% to 12.8% of primary cardiac tumors (4, 6). The most frequent are synovial sarcoma (approximately 5% of cardiac sarcoma),angiosarcoma, and undifferentiated pleomorphic sarcoma (1). According to our center data of ten years, six cases were diagnosed as pericardial malignancies. Three were synovial sarcomas, including two males (7 years old and 31 years old) and one female (50 years old). It coincides with a previously reported trend that pericardial synovial sarcomas are more common in young men (3, 7).

Most of the patients were hospitalized because of dyspnea, cough, chest pain, edema of lower limbs, and other symptoms caused by heart failure caused by pericardial effusion or tumor oppressing the heart (8). Echocardiography and chest CT are often used in diagnosis; however, a cardiac MRI may help for the surgery approach (3). More than 90% of synovial sarcomas contain a chromosomal translocation in t(X;18)(p11.2;q11.2), and it would be helpful in diagnosis (9-11). Due to these diseases' rare occurrence,

this tumor's treatment is unidentified, but most patients get surgical resection. Although pooled analysis performed that surgical treatment was no impact on overall survival,two—thirds of patients who do not get surgery died within the first six months. The researcher believed it was due to the frequent failure of complete resection, complete resection is recommended as it was the only independent prognostic factor associated with survival. (3) Postoperative adjuvant therapy is also controversial; consider incomplete resection and the risk of metastases, Ganesh Shanmugam recommended using adjuvant chemotherapy and radiotherapy (12). But radiotherapy is not the preferred method because of cardiac complications in a long time follow-up (13, 14). The prognosis is recognized to be miserable despite the treatment, and the 5-year survival rate of pericardial synovial sarcoma was 22.2% (15), the median survival was 27 months (3), maximum survival is 14 years (16).

For the safety of our patient, we used the following methods in the operation: 1. To control the operation process and avoid massive bleeding and heart damage, we used the segmented resection method; 2. Prepare extracorporeal circulation to deal with emergencies.

In our case, we noticed that the patient began to have symptoms four months before admission and had been examined in two other hospitals with echocardiography. Still, only pericardial effusion was found, but no tumor. The tumor wasn't discovered until four months after the onset of the symptoms. In the literature, we found the same cases with pericardial effusion but no tumor on echocardiography. And like our case, they had a fever as the first symptoms (17, 18). We recommend CT be performed in patients with recurrent fever and sizeable pericardial effusion to rule out possible pericardial synovial sarcoma considering the echocardiography limitations.

List of abbreviations:

CT: Computed tomography

MRI: magnetic resonance imaging

PSS: Pericardial synovial sarcoma

PET/CT: positron emission tomography

Authors' contributions

QL and ZH collected data; QL analysed the patient data and wrote the first draft of this manuscript. ZH and SG helped to draft the manuscript and revised it critically for important intellectual content. All authors read and approved the final manuscript.

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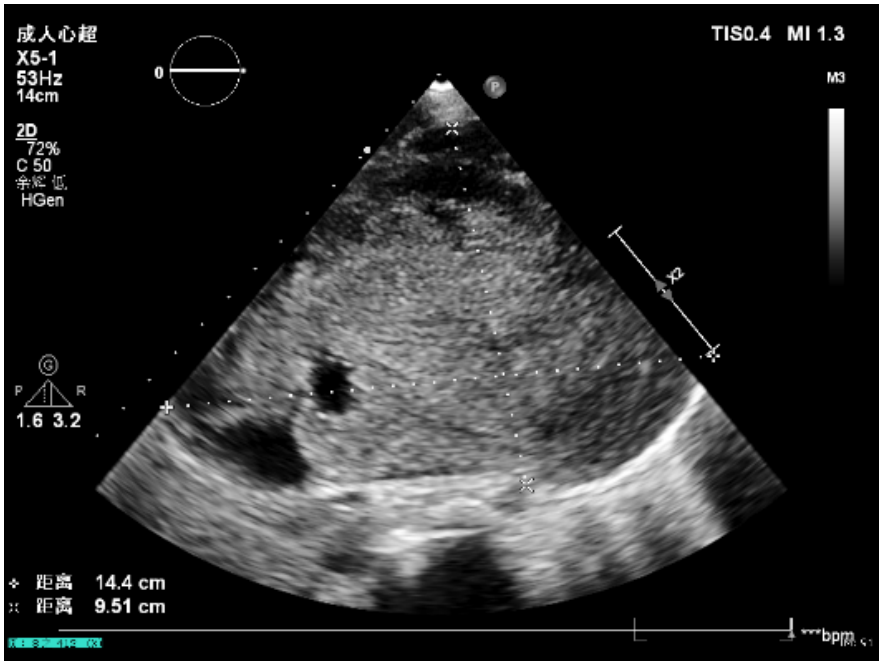
Figure Legends

Figures



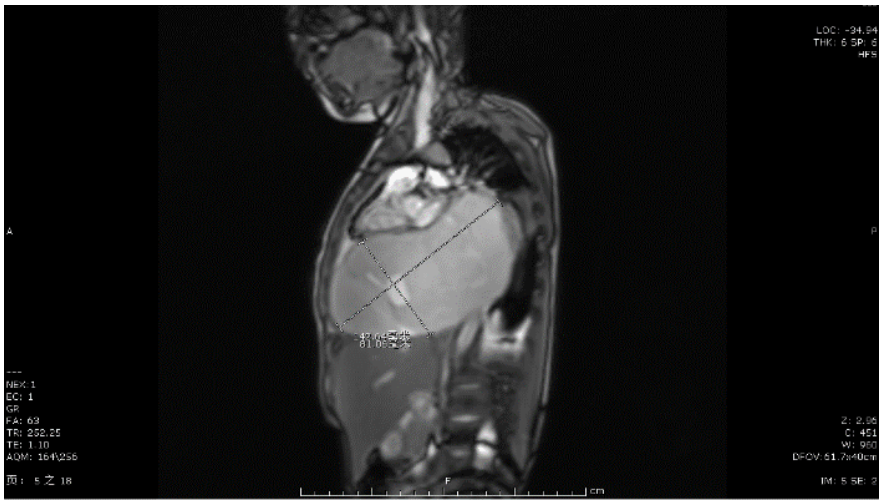
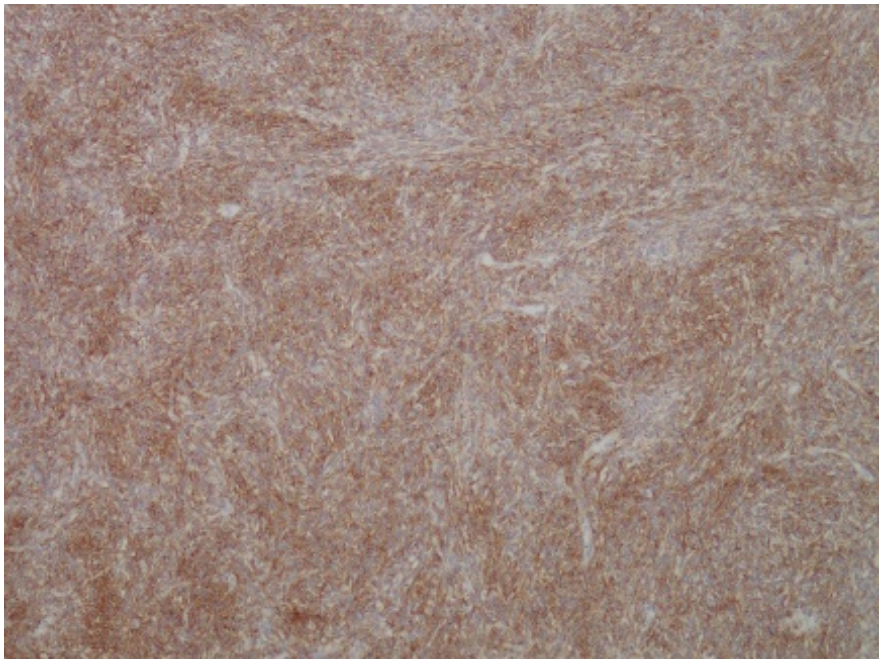
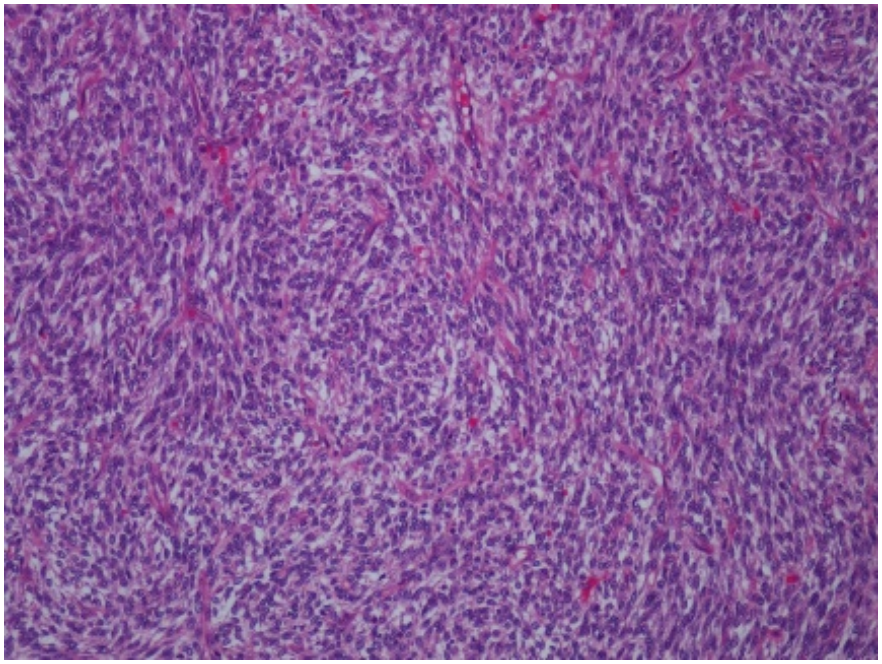
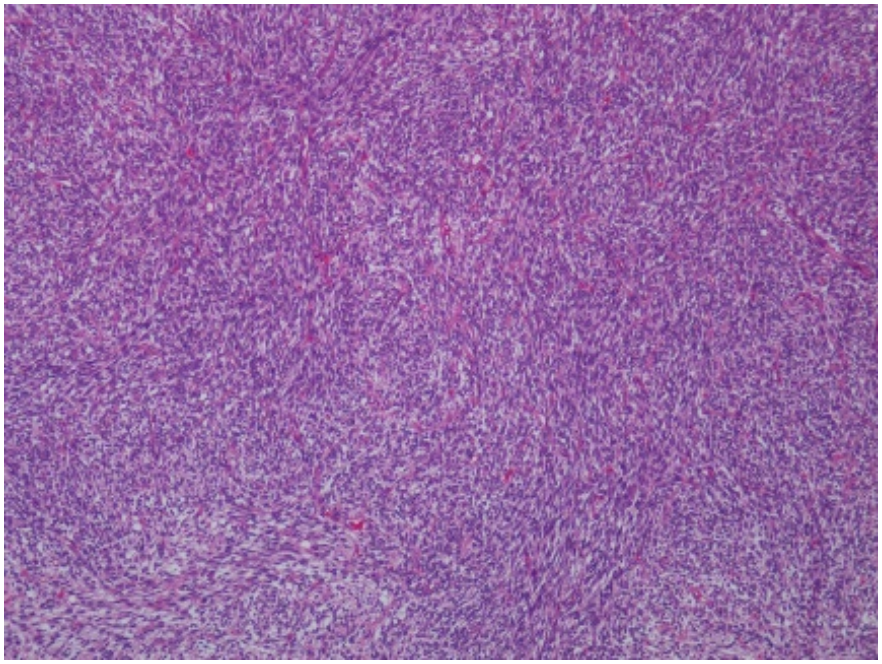


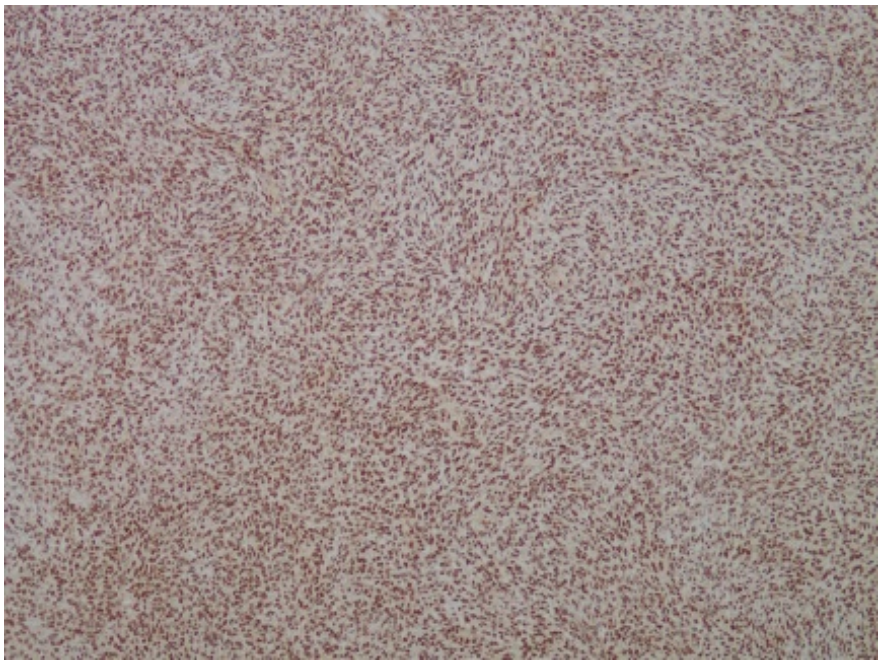
Figure 1(A) Figure 1(B)

Figure 1: A: A huge irregular hypoechogenic area with a range of about 144 × 95 mm was seen in the pericardial cavity, and an irregular echogenic area was seen. B:A cardiac Magnetic Resonance Imaging (MRI) demonstrated a 142×94×81-mm mass below the heart in the pericardial cavity and close to left and right ventricles and inferior wall of left and right atria.









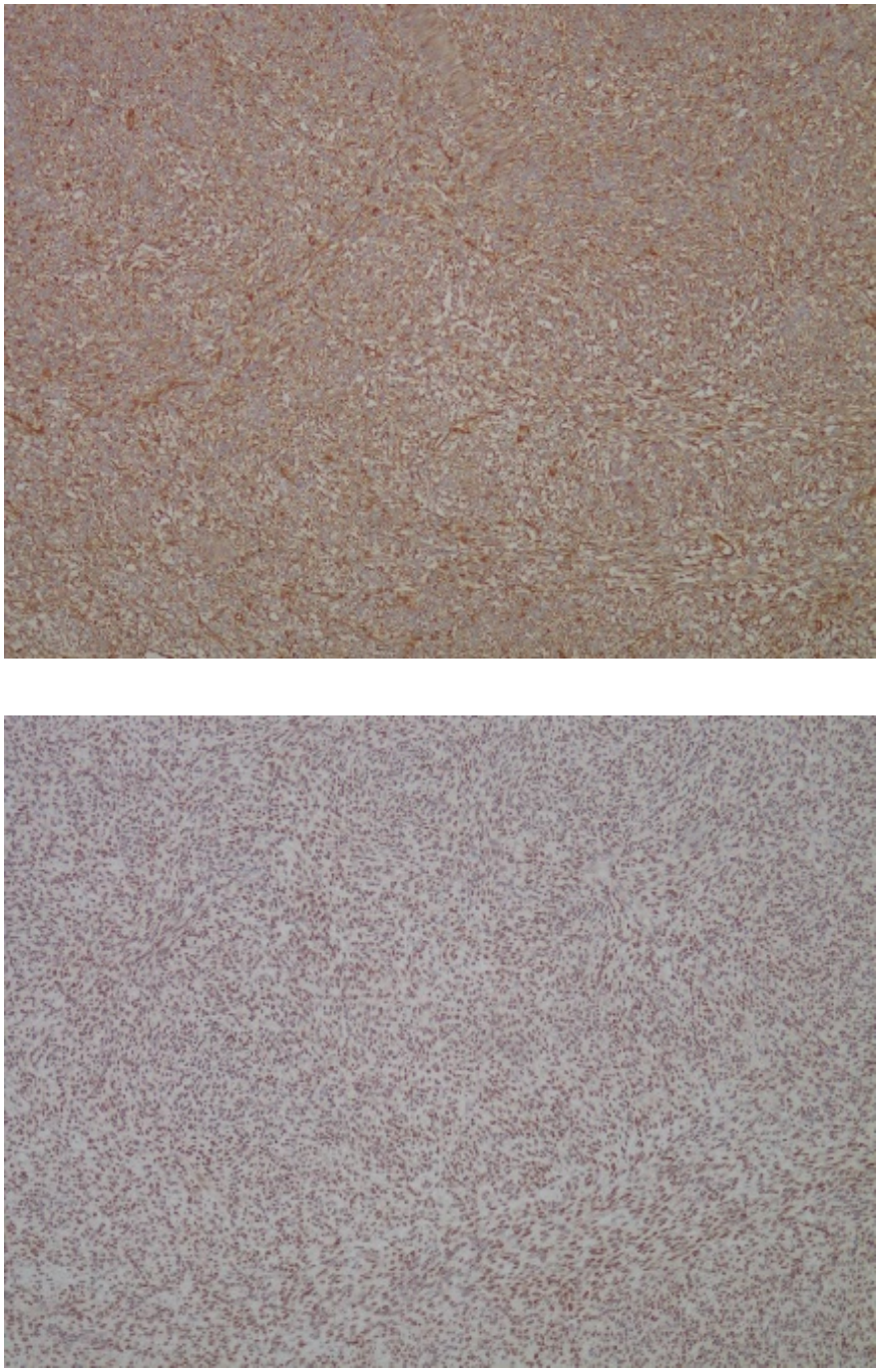


Figure 2(A) Figure 2(B) Figure 2(C)

Figure 2(D) Figure 2(E) Figure 2(F)

Figure 2: The histopathological and immunohistochemical finding of tumor tissue. Photomicrograph shows monophasic synovial sarcoma in which spindle cells Hematoxylin and Eosin staining ×100(A), ×400(B). The cells were positive for EMA(C), INI1(D), TLE-1(E), VIM(F). Magnification ×100.

Declarations

Ethics approval and consent to participate

The case only involved objective retrospective descriptions, so ethics approval was not required. The patient provided consent for the use of the physical and imaging information adopted from the patient.

Consent for publication

Written informed consent was obtained from the patient's parent for publication of this case report and any accompanying images. The consent form will be provided upon request.

Availability of data and materials

This case report only contains clinical data from the medical records of the patient reported herein. The data will be made available upon request.

Competing interests

The authors declare that they have no competing interests.

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