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CASE REPORT

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Inflammatory myofibroblastic tumor of the pericardium in an 11-month-old infant: A case report

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Abstract

Primary cardiac tumors are very rare in infants. Here we present an 11-month-old infant with a pericardial inflammatory myofibroblastic tumor who presented with symptoms of respiratory distress and cardiac tamponade. The tumor was surgically removed, and the patient received medical treatment; the patient had no problem with follow-up.

KEYWORDS

cardiac tumor, inflammatory myofibroblastic tumor, pediatrics, pericardium

1 | INTRODUCTION

Primary cardiac tumors are very rare in infants (0.13% incidence). Most are benign lesions such as rhabdomyomas, fibromas, teratomas, or lipomas.[1] Inflammatory myofibroblastic tumor (IMT), classified as either a neoplasm or an inflammatory tumor, constitutes less than 5% of all primary heart tumors.[2] IMT is usually benign but has the potential to be recurrent, invasive, and malignant. Clinical presentation, depending on the organs affected, varies from asymptomatic to sudden death as a result of cardiogenic syncope or myocardial infarction owing to embolization of the tumor into the coronary arteries.[3] This report describes an 11-month-old infant with a pericardial inflammatory myofibroblastic tumor who presented with symptoms of respiratory distress.

2 | CASE PRESENTATION

An 11-month-old girl who was previously healthy has been referred to a children's hospital with symptoms of respiratory distress and a history of upper respiratory infection (URI) symptoms for 2 weeks. Pericardial tamponade was

initially suspected due to the muffled heart sounds and weak pulses.

A chest X-ray revealed cardiomegaly and a large opacity on the left side, causing a mediastinal shift. Additional echocardiographic examination confirmed the diagnosis of a pericardial mass adhering to the left atrial wall and tamponade (Figure 1). Pericardiocentesis was done immediately due to critical tamponade symptoms.

After taking a computed tomography (CT) scan (Figure 2) and doing the necessary surgical consultations, the patient was referred to a tertiary heart hospital for surgery.

The tumor was completely removed in surgery on a beating, perfused heart. A 4×6 cm tumor that originated from the left atrial appendage and extended to the whole posterior and left pericardial space with no invasion to the heart or other adjacent tissues was completely removed.

Macroscopic pathological examination revealed a semioval-shaped, capsulated, creamy mass measuring 6×4×2 cm that showed some hemorrhagic areas (Figure 3), and microscopic pathological examination revealed a cellular neoplasm composed of myofibroblastic and fibroblastic spindle cells with inflammatory cells including lymphocytes, plasma cells, eosinophils, neutrophils, and histiocytes arranged in a fascicular pattern in abundant blood vessels in the background.

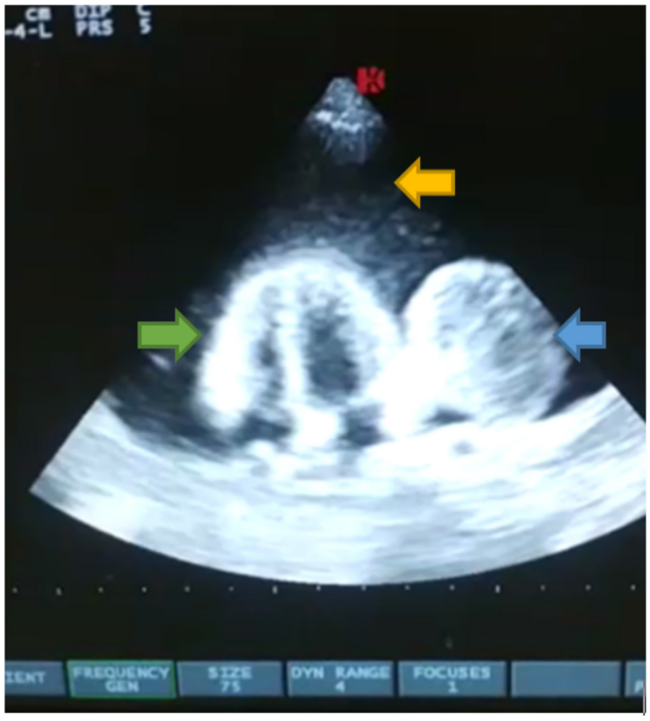

FIGURE 1 An echocardiogram revealed a massive pericardial effusion with large mass: blue arrow: mass, yellow arrow: massive pericardial effusion, green arrow: heart.

In the immunohistochemistry study, vimentin and smooth muscle actin (SMA) were positive, and pan cytokeratin antibody (PCA) and calretinin were negative.

According to pathological results, the diagnosis is an inflammatory myofibroblastic tumor. In the chest CT scan and cardiac magnetic resonance image (MRI), which were taken 2 and 6 months after tumor resection, there was no residual mass (Figure 4).

After 1.5 years of follow-up, the patient's condition was appropriate, and there was no recurrence or problem.

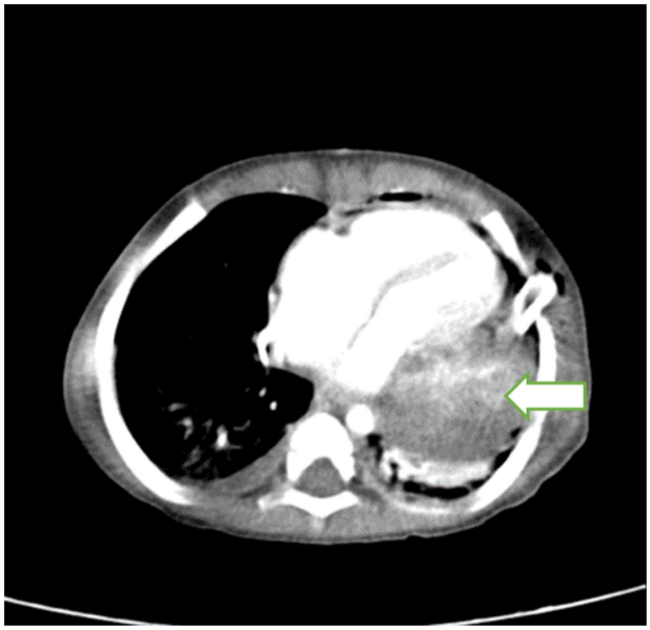

FIGURE 2 Chest CT scan after pericardial drainage: arrow: mass.

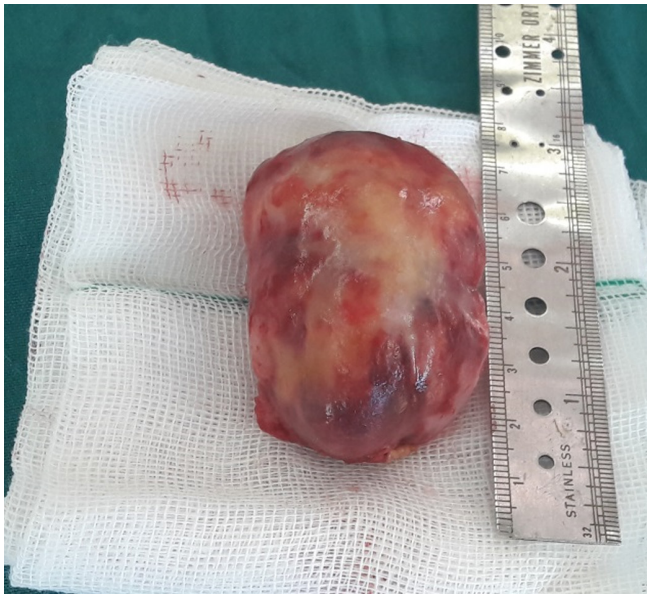

FIGURE 3 Macroscopic view of the tumor.

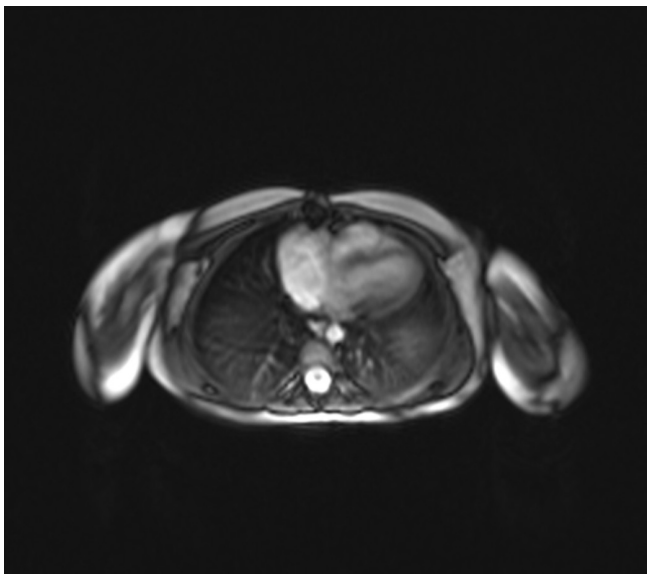

FIGURE 4 Cardiac MRI 6 months after tumor resection that showed no residual mass.

3 | DISCUSSION

Inflammatory myofibroblastic tumors, which have a predilection for the lung, abdomen, and pelvis, are rare, benign primary tumors of the heart in infants.[4,5] The IMT etiology remains unknown and controversial. Due to the prominent inflammatory infiltrate and associated systemic symptoms in a minority of patients with IMT, a viral etiology (such as HHV-8 or EBV) has been proposed; other potential causes involve the response to previous injury and chromosomal aberration of region 23 within the short arm of chromosome 2.[5] Rearrangements involving the ALK locus on chromosome 2p23 have been documented in approximately 50% of IMTs. Distant metastases occur primarily in ALK-negative IMTs, but local recurrence occurs regardless of ALK expression. Several ALK fusion proteins, including TPM3-ALK found in IMT, induce transformation in cell lines and animal models, a finding that suggests that ALK rearrangement may define a subgroup of IMTs that is sensitive to targeted kinase inhibition such as ALK inhibitor crizotinib (PF-02341066, Pfizer).[6] In this patient, we did not study gene analysis due to family circumstances and facilities. The right atrium, right ventricle, tricuspid valve, and interventricular septum are the primary sites of cardiac involvement in IMT.[2–8] patients with IMT generally present with nonspecific symptoms, including vague abdominal pain or gastrointestinal complaints for intraabdominal lesions and cough, chest pain, or, less often, hemoptysis for pulmonary tumors. A constitutional syndrome consisting of fever, weight loss, and malaise is seen in 15–30% of patients, and laboratory evaluation may reveal microcytic anemia, a raised erythrocyte sedimentation rate, thrombocytosis, and/or polyclonal hypergammaglobulinemia.[5] our patient had a history of URI with symptoms of mild fever, cough ad sore throat for 2 weeks.

IMTs range from 1 cm to >20 cm in greatest dimension, with a mean size of 6 cm.[5] IMTs are classified as tumors of intermediate biological potential by the most recent World Health Organization classification, due to a tendency for local recurrence and a low risk of distant metastasis.[7] The recurrence rate varies by anatomical site, from 2% for tumors confined to the lung, to 25% for extrapulmonary lesions.[9] Clinical, laboratory, radiological, and histopathological confirmation are required for the diagnosis.

Inflammatory myofibroblastic tumor has the potential to undergo malignant transformation or metastasize. Surgical removal of the tumor for diagnosis and treatment should be the mainstay of management. There is no medical therapy indicated as the primary therapy for IMTs. Scattered reports on the effectiveness of Celecoxib, a nonsteroidal anti-inflammatory drug (NSAID), were available. Combination of Celecoxib with chemotherapeutic agent was reported to induce durable remission.[10] In some cases, especially when the tumor is aggressive owing to metastasized lesions or when the tumor is not resectable, chemotherapeutic agents (such as methotrexate, vinblastine, vinorelbine, vincristine, cyclophosphamide, actinomycin-D, and ifosfamide-based chemotherapy) and/or radiotherapy may be required.[11] It is well known that IMTs respond to corticosteroid therapy. Steroids, on the other hand, have been reported to hasten the progression of IMTs.[12] The optimal management of locally aggressive and metastatic forms is still under debate and should be decided on a case-by-case basis.

In special cases, cardiac surgical intervention such as valve replacement and valvular or vascular reconstruction may be required, and polytetrafluoroethylene grafts are used for bypass grafting. When the tumor is aggressive or there is recurrence in the heart, transplantation may be considered.[13] Based on a good surgical resection and normal post-resection CT and MRI results, our patient received only ASA and corticosteroids. Oral prednisolone was administered at a dose of 2 mg/kg/day for 2 weeks, then tapered and discontinued. Now she is in good condition without problems after more than 1.5 years of follow-up. We planned to follow-up for recurrences by echocardiography and testing for thrombosis and C-reactive protein as an inflammatory marker until adulthood.

4 | CONCLUSION

Cardiac IMTs are usually benign lesions and may be the cause of the enhanced cardiovascular and respiratory

dysfunction in infants. It is typically located within the endocardium but can also develop within the pericardium. As cardiac IMT can be potentially fatal, early clinical diagnosis is critical. Tumor resection is a treatment of choice. Steroid therapy, or chemotherapy, is a medical alternative to surgery if tumor resection is not feasible. Patients with IMTs after surgery should be informed about the nature of this entity and must be followed up with echocardiography and specified hemochemical examinations.

AUTHOR CONTRIBUTIONS

All authors have substantially contributed to this work. MR Khalilian, BSH Shamsian, R Baghaei Tehrani, L Kharaz, and N Talebian: gave substantial contributions to the conception and design of the work; drafted the work and critically revised it for important intellectual content; and gave final approval of the version to be published.

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FUNDING INFORMATION

None to declare.

CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

This case was approved by the Mofid Children's Medical Research Center. The parents of the patient gave written informed consent to publish the case.

CONSENT

Written informed consent was obtained from the participant for the publication of the details of their medical case and any accompanying images.

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