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Clinical Case Report

Rare presentation of four primary pediatric cardiac tumors

Lauren R. Schaffer [a], Shelley A. Caltharp [a], Sarah S. Milla [b], Brian F. Kogon [c], Caitlin A. Cundiff [a], Aarti Dalal [d], Phillip C. Quigley [a], Bahig M. Shehata [a,*]

[a] Department of Pathology, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, GA, US
[b] Department of Radiology, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, GA, USA
[c] Department of Pediatric Cardiac Surgery, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, GA, USA
[d] Department of Pediatric Cardiology, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, GA, USA

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ABSTRACT

Pediatric cardiac tumors are extremely rare and usually benign. We selected four unique cases of pediatric cardiac tumors from a 15-year period at our institution. The four chosen cases represent unique, rare primary tumors of the heart. Our selection includes a case of Rosai Dorfman disease without systemic involvement, which is, to our knowledge, the second case of isolated cardiac Rosai Dorfman disease in a child. We present a case of subtotal replacement of myocardium by granulocytic sarcoma with minimal bone marrow involvement, representing the first reported case in a child manifested as hypertrophic cardiomyopathy, as well as a case of a primary synovial sarcoma arising from the atrioventricular (AV) node, representing the fourth reported pediatric case of a cardiac synovial sarcoma, and it is the first to arise from the AV node. Finally, we present a primary congenital infantile fibrosarcoma of the heart, which is, to our knowledge, the first confirmed cardiac congenital infantile fibrosarcoma. These four cases represent the need for continued inclusion of rare cardiac conditions in a clinician's differential diagnosis. Furthermore, they present the need for more in-depth molecular and genomic analysis of pediatric cardiac tumors in order to identify their etiopathogenesis.

1. Introduction

Cardiac tumors are uncommon, as evidenced by an autopsy incidence rate of 0.001% to 0.03% [1]. While primary benign cardiac tumors are rare, primary malignant tumors are even less common, particularly in children — only 10% of pediatric cardiac tumors are malignant compared to 25% of the cardiac tumors found in the adult population. The most common pediatric cardiac tumors are rhabdomyoma (40–60%), fibroma (12–16%), teratoma (15–19%), and myxoma (2–4%), all of which are benign. In terms of malignancies, sarcomas represent 2% of all pediatric cardiac tumors, with angiosarcomas and rhabdomyosarcomas the most common [2,3]. When diagnosed with a malignant cardiac tumor, the prognosis is usually poor because the tumor has often metastasized to other organs by that time [1]. Although cardiac tumors are rare, clinicians should include them in their differential diagnosis when a patient presents with a murmur, arrhythmia, or heart failure symptoms.

2. Case presentations

We searched our files from 2000 through 2014 for cases of cardiac tumors. We found 27 cases — 19 of the tumors were benign, 6 were malignant, and 2 were metastatic. Of those cases, we present 4 that are unusual and exceptionally rare (Table 1). The other cases represent myxomas (8), rhabdomyomas (7), teratomas (5), fibromas (2), and metastatic neuroblastoma (1).

2.1. Case 1

A 12-year-old male with sickle cell disease presented with chest pain and fever. The use of telemetry revealed that he had a complete heart block prompting further investigation. An echocardiogram identified an intracardiac mass originating above the tricuspid valve and extending into the interatrial septum, as well as encircling the aortic root (Fig. 1). A needle biopsy was performed, and the histology demonstrated the distinctive finding of emperipolesis. The tissue-staining patterns were also strong for S-100. This evidence led to the diagnosis of Rosai Dorfman disease (RDD) [4]. Uniquely, there was no lymph node or other organ involvement. The patient required a dual chamber pacemaker and, eventually, underwent cardiac resynchronization therapy (CRT). However, his heart's function continued to decline until he had a successful orthotopic heart transplant. The explanted heart showed cardiomegaly, weighing 415 g (expected weight, 232 g). The mass appeared to be arising from interatrial septum, occupying the right atrium

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* Corresponding author at: Atlanta, GA 30329, United States. Tel.: +1-404-785-1390.
E-mail address: bshehat@emory.edu (B.M. Shehata).

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Table 1
Clinical Pathological Description of 4 Rare Primary Pediatric Cardiac Tumors

Case	Age/Sex	Ethnicity	Clinical presentation	Pathological	Diagnosis	Treatment	Survival/Follow-up
1	12/M	African American	Chest pain, fever	Emperipolesis, positive S-100 stain	Isolated cardiac RDD	Needle biopsy; dual chamber pacemaker; CRT; heart transplant	Surviving posttransplant 18 months later
2	7/M	Caucasian	Tachypnea, shortness of breath, nonspecific anemia	Myocardial infiltration by myeloid precursors; myocyte necrosis	AML	Bone marrow biopsy; ECMO	Expired
3	13/M	Caucasian	Flu-like symptoms	Biphasic; spindle and epithelial components; cell junctions, interdigitating cell membranes, intercellular lumina with microvilli	Primary cardiac synovial sarcoma	Resuscitation in ER failed	Expired (cardiac arrest)
4	7 months/M	Caucasian	Lethargy, bloody stools	Spindle cell neoplasm, staghorn blood vessels	CIFS of the heart	Pericardiocentesis; open biopsy; chemotherapy; surgery to repair ventricle rupture	Surviving 6 years

and infiltrating the left ventricle and aorta. Histological examination of the mass showed lymphoplasmocytic infiltrate with occasional histiocytes engulfing multiple inflammatory cells (emperipolesis). The histiocytic cells stained strongly positive for S-100 (Fig. 2).

2.2. Case 2

A 7-year-old male presented with respiratory distress. He had previously been seen multiple times as an outpatient for nonspecific anemia and thrombocytopenia. During those visits, he had an extensive outpatient oncologic workup, including two negative bone marrow biopsies. It was postulated that his bone marrow suppression was due to a viral illness. However, when he presented in the clinic 1 month later with shortness of breath and tachypnea, he was sent to the emergency department for a chest X-ray. It showed cardiomegaly, which prompted an echocardiogram. The echocardiogram confirmed the cardiomegaly and revealed biventricular hypertrophy (Fig. 3A). Hematological workup, including a bone marrow biopsy, revealed acute myeloblastic

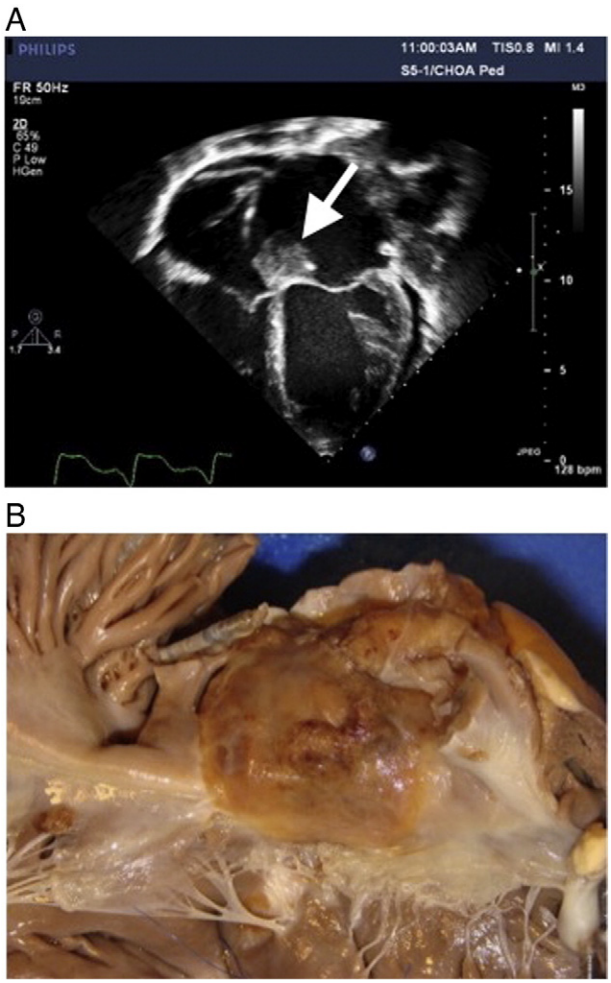

Fig. 1. Case 1, echocardiogram showing the mass originating above the tricuspid valve and extending into interatrial septum. (A) Case 1, tumor originating above tricuspid valve and invading the interatrial septum.

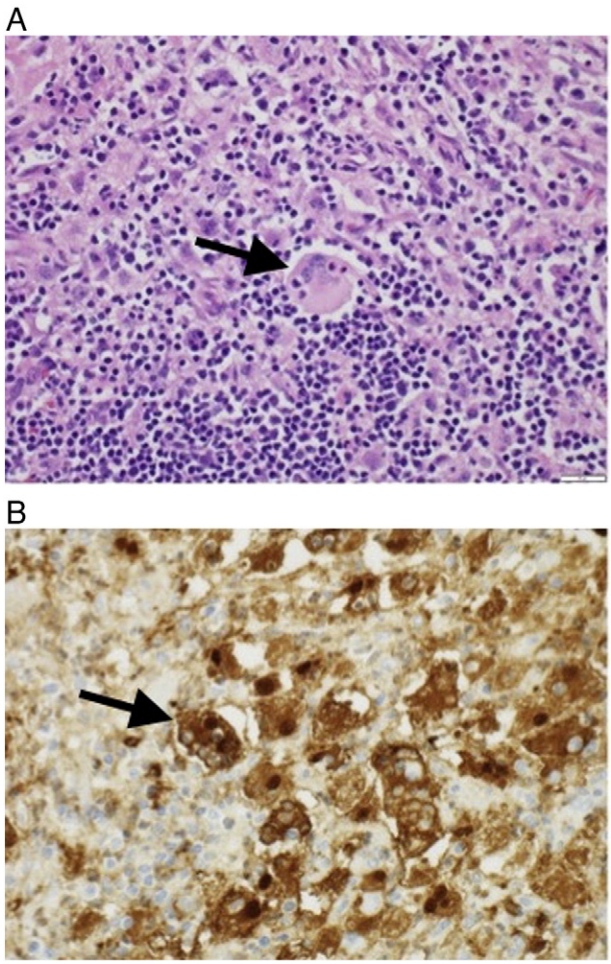

Fig. 2. (A) Case 1, H&E stain (400×) showing histiocytes engulfing lymphocytes and erythrocytes (emperipolesis) (see arrow). (B) Case 1, S-100 immunostain (400×) showing strong positivity of the histiocytes.