We report a case of a child with a right ventricular inflammatory myofibroblastic tumor (IMT) who presented with fever, viral symptoms, and abdominal discomfort. Including this case, 49 intracardiac tumors have been previously reported in all age groups. The majority of intracardiac IMTs occur in pediatric patients, with approximately half presenting in children aged <12 months. Intracardiac IMTs are generally described as benign tumors; however, depending on their location, the initial presentation may involve heart failure or sudden death. In addition to cardiac signs and symptoms, the clinical presentation of IMTs may also include constitutional signs such as fever, anemia, and elevated inflammatory markers. This case report reviews the diagnosis and management of IMTs, as well as the histopathologic features of this rare tumor type. Clinicians should be aware of their clinical presentation because early diagnosis and treatment can significantly reduce morbidity and mortality.

**abstract**

Inflammatory myofibroblastic tumors (IMT) are rare types of spindle-cell tumors that are characterized by myofibroblastic lesions with lymphoplasmacytic infiltrates. IMTs are rare among the cardiac tumors. Including our case, 49 cases of intracardiac IMTs have been reported across all age groups (see Supplemental Information). Of these, 33 have been reported in the pediatric population, with approximately half of this group involving children <12 months of age. Although IMTs are generally described as benign tumors, a small number demonstrate neoplastic properties, such as local recurrence after resection.

The clinical presentation of IMTs often includes fever, respiratory distress, anemia, weight loss, and elevated inflammatory markers. Patients may also first present with cardiac signs and symptoms such as respiratory distress or sudden death. Deaths due to the tumor are related to its location in the heart; lesions located near the coronary arteries, cardiac valves, and ventricular outflow tracts, in particular, are associated with fatality.

**CLINICAL RECORD**

An 11-month-old previously healthy female presented to the emergency department with a 5-day history of intermittent fever and irritability. There was no history of diarrhea, bloody stools, or sick contacts. Her medical history was otherwise unremarkable. Growth parameters were appropriate for age. Family history was positive for sickle-cell trait and negative for congenital heart disease. She was brought to a walk-in clinic on 2 occasions and discharged from the hospital with a provisional diagnosis of viral illness. Her symptoms continued to worsen over time, and she was brought to the hospital emergently. On examination, she was febrile and tachypneic. Her heart rate was 200 beats per minute, and her blood pressure was 60/30 mm Hg. A chest radiograph showed cardiomegaly with a pleural effusion. An echocardiogram revealed a mass in the right ventricle, with severe tricuspid regurgitation. She was taken to the operating room for emergent surgical resection of the tumor. The postoperative course was uneventful, and she was discharged home on postoperative day 3.

the next 24 hours as she developed decreased energy, a mild cough, and a single episode of nonbilious emesis.

Presenting vital signs were significant for mild tachycardia and tachypnea, and her axillary temperature was 101.7°F. Examination revealed a clinically stable child who was fairly settled. She had a grade 3 systolic crescendo-decrescendo murmur at the left lower sternal border. She had some subternal retractions, but her lung fields were clear on auscultation. Her abdomen was significantly distended and tender on examination. Liver edge was 2 cm below the right costal margin. The remainder of her examination was normal. Laboratory investigations revealed a white blood cell count 22,000 cells/μL, with a predominance of neutrophils (13,400 cells/μL), hemoglobin 8.1 g/dL, and platelets 469,000 cells/μL. C-reactive protein was elevated at 12.3 mg/dL. She had a compensated lactic acidosis. Chest film revealed a cardiothoracic ratio of 0.55, the upper limit of normal, but lung fields were clear. An abdominal ultrasound revealed mild hepatic enlargement, free fluid, and a small right pleural effusion. The patient was admitted to pediatric general surgery for further investigation and management of a presumed gastrointestinal process and treated empirically with antibiotics. Given the new murmur, cardiology was consulted. The patient remained stable overnight.

On postadmission day 1, her tachypnea progressed from a respiratory rate of 30 to 50 breaths per minute on room air while maintaining oxygen saturation of 100%. Her respiratory status continued to deteriorate over the course of a few hours as she developed progression of her cough, grunting, and perioral cyanosis. She was transferred to the ICU for respiratory support and monitoring, and cardiology was reconsulted on an urgent basis. She required active resuscitation and inotropic support to maintain perfusion. An echocardiogram performed during the resuscitation revealed a right ventricular mass obstructing her right ventricular outflow tract, significant right ventricular hypertrophy, and depressed left ventricular filling (Fig 1). The patient developed a narrow complex bradycardia and eventual cardiac arrest. The patient died during resuscitation attempts.

At autopsy, cardiac evaluation revealed a heart that weighed 65.5 g (normal range 49 ± 6 g). The right ventricle width measured 0.4 cm and was severely dilated. There was a visible bulge on the anterior basal area of the heart corresponding to a yellow-red polypoid tumor, measuring 2.7 × 2.2 × 1.5 cm, arising from the posterior wall of the right ventricle just below the pulmonary valve (Fig 2A). Both atria were dilated. No other abnormalities were noted in the cardiac structure. On examination of the body cavities, bilateral pleural effusions (right 30 mL, left 100 mL), hydropericardium (30 mL), and ascites (300 mL) were noted. Postmortem blood, cerebrospinal fluid, and pleural fluid cultures were negative. A postmortem nasopharyngeal swab for respiratory viruses was also negative.

Histologic examination revealed a spindle-cell lesion with scattered mitoses of up to 2 mitoses per 10 high power field (Fig 2B). There was no invasion of the myocardium. There were occasional foci of mononuclear inflammatory cells, and multiple areas of focal necrosis with overlying thrombi. The tumor stained positive for vimentin, supporting a mesenchymal origin. The spindle-cell component of the lesion stained positive for muscle-specific actin and smooth muscle actin, consistent with a myofibroma (Fig 2C). Desmin (various muscle types) and myogenin (skeletal muscle) both stained negative. The tumor also stained negative for anaplastic lymphoma-kinase-1 (ALK-1). The tumor stained negative for other cell lineages, including S100 and cytokeratin AE1/ AE3 (CAE1/AE3), therefore negative for neurogenic and epithelial markers, respectively. The final diagnosis of IMT was made on the basis of the presence of inflammatory cells with positive staining for muscle-specific actin and smooth muscle actin.

**DISCUSSION**

Cardiac tumors are rare in the pediatric population. The incidence in both adults and children is between 0.0017% and 0.028%. Rhabdomyomas are the most common cardiac tumor in children >12 months of age, encompassing >60% of all childhood cardiac tumors, followed by fibromas.
and myxomas.\textsuperscript{7,8} Although intracardiac IMTs are rare, they have a predilection for younger patients. Including this case, only 49 intracardiac IMTs have been reported worldwide; of these, \textgreater \textasciitilde two-thirds have appeared in patients under age 18 years (33 of 42), with about half of them appearing under age 12 months (17 of 33).\textsuperscript{1,3-5,8}

The differential diagnosis for intracardiac IMTs include some of the more common cardiac tumors, particularly rhabdomyomas, myxomas, and fibromas. These are differentiated from IMTs based on histology and extent of invasion into the myocardium.\textsuperscript{9,10} IMTs appear histologically similar to myosarcomas but can be differentiated on the basis of their having less cellular pleomorphism, atypia, and fewer mitotic figures\textsuperscript{1,10} and not invading into the myocardium. ALK-1 is positive in 35\% of IMTs,\textsuperscript{11} although a negative result does not rule out the diagnosis because several intracardiac IMTs have previously stained negative.\textsuperscript{2,8,12,13} IMTs are described as benign reactive lesions; although the exact etiology of the tumor is unknown, several studies have associated IMTs with Epstein-Barr infection.\textsuperscript{8,11-13} 

Local recurrence is reported in up to \textasciitilde 10\% of cases.\textsuperscript{4} Recent research has also suggested that IMTs may exhibit some chromosomal aberrations at the 2p23 locus, further supporting the possibility of a neoplastic mechanism of disease.\textsuperscript{11}

Intracardiac IMTs have a variable clinical presentation depending on the site of the tumor. They are also usually associated with constitutional signs and symptoms including fever, anemia, polyarthritis, and vascular compromise, which are thought to be caused by the release of cytokines by IMTs, interleukin-6, in particular.\textsuperscript{5,11} They can also present with dyspnea, which may manifest as respiratory distress in infants that should be monitored and investigated.

Young infants and children in heart failure may appear otherwise well during the earlier stages because their cardiovascular physiology compensates significantly to maintain cardiac output. However, decompensation is rapid if the underlying cause is not addressed. The location of the tumor for our patient was challenging for stabilization because the mass was obstructing the right outflow tract, limiting blood delivery to the pulmonary and systemic circulation systems. Rates of sudden death due to cardiac tumors is \textasciitilde 0.06\% in persons 34 years and younger.\textsuperscript{15} Poor prognosis is associated with lesions involving the coronaries, cardiac valves, or ventricular outflow tracts.\textsuperscript{2,5}

Although serum laboratory investigations may offer some information, diagnostic imaging is the mainstay for diagnosis of cardiac tumors. An echocardiogram is the most useful modality to confirm diagnosis of an intracardiac mass.\textsuperscript{16} The differential diagnosis for an intracardiac mass is included in Table 1. Although echocardiography can be helpful to differentiate the etiology of the lesion,\textsuperscript{16} histologic examination remains the gold standard for confirmation of the diagnosis.

IMTs are considered a benign tumor. Intracardiac IMTs are treated definitively by completely resecting the tumor. The prognosis after excision is good, although there is
For unresectable tumors, heart transplantation may be considered as an option, although there has been only 1 successful case reported for this indication. Corticosteroids have been used previously as adjunct therapy for difficult resections and for IMTs in other body sites. There is no current evidence that they prolong survival for intracardiac tumors in particular. Therefore, surgical resection remains the mainstay of treatment.

**CONCLUSIONS**

Inflammatory myofibroblastic tumors may present with constitutional symptoms that can mimic other common illnesses. Congestive heart failure and obstructive shock are end-stage clinical presentations that require urgent echocardiography and imaging for diagnosis and management. Laboratory investigations can provide ancillary information about perfusion and oxygenation. A high index of suspicion is necessary to make the diagnosis of this rare lesion because timely surgical resection is ultimately required for the definitive and potentially lifesaving management.

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**ABBREVIATION**

IMT: inflammatory myofibroblastic tumor

**REFERENCES**


Febrile Illness in an Infant With an Intracardiac Inflammatory Myofibroblastic Tumor
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*Pediatrics* originally published online January 21, 2016;

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