A previously healthy 15-year-old boy was admitted after 6 months of intermittent fevers, night sweats, and 9 kg weight loss. A systolic murmur was appreciated at the left upper sternal border. Laboratory data revealed elevated white blood cells (11 800/µL), erythrocyte sedimentation rate (75 mm/h), and C-reactive protein (10.5 mg/dL). The results of blood cultures were negative. Imaging demonstrated a hypermetabolic 2×2×4.5 cm pedunculated mass in the right ventricle, suggestive of a neoplasm with overlying thrombus (Figures 1A, 2A and 2B and online-only Data Supplement Movie I).

At surgery, the mass was found to originate from the midportion of the interventricular septum and was covered with thrombus. Debridement revealed a firm whitish mass infiltrating the septum and involving the base of several tricuspid valve (TV) attachments. Intraoperative frozen sections revealed only thrombus and infarct. A subtotal resection was performed to avoid resection of the septum and TV in the absence of a diagnosis of malignancy. Final pathology revealed whorled fibrous tissue with myofibroblasts, patchy inflammation, and acute and organizing thrombus consistent with a diagnosis of inflammatory myofibroblastic tumor (IMT) of the heart (Figure 3A). The results of tissue cultures, autoimmune, and hypercoagulable workups were negative.

The patient’s symptoms resolved, and there was no evidence of recurrence on surveillance echocardiography (Figure 2C and 2D) until 2 years later when he again experienced fevers. Imaging revealed a 2.8×3.8×5.2 cm elongated mass arising from the right ventricular free wall below the anterior leaflet of the TV (Figure 1B and online-only Data Supplement Movie II).

Surgical resection was considered high risk given the need for repeat sternotomy and the proximity of the tumor to the TV and right main coronary artery. A trial of prednisone was attempted given reports of IMT regression with corticosteroids.1 The patient’s symptoms resolved with steroid treatment and tumor size remained stable. He was tapered off of prednisone after 6 months of treatment but again experienced fevers. Imaging revealed tumor enlargement to 3.5×4.0×5.0 cm with involvement of the anterior and posterior leaflets of the TV, impeding leaflet mobility and coaptation (Figure 4A and 4B). The patient was taken for repeat resection given tumor enlargement and compromised TV function. At reoperation, all 3 leaflets of the TV were heavily involved and were removed with the friable tumor in piecemeal fashion. After resection of all gross tumor, a 27-mm bioprosthetic valve was placed. Intraoperative and immediate postoperative echocardiography revealed no evidence of residual tumor. Pathology showed recurrent IMT (Figure 3B). The patient was placed on daily ibuprofen given reports of IMT responsive to nonsteroidal anti-inflammatory medications2 and the lack of evidence supporting chemotherapy or radiation after complete resection.1

Over the next month the patient continued to experience fevers, and C-reactive protein level increased from 4.6 to 16.6 mg/dL. Echocardiography revealed an increase in the mean pressure gradient across the TV with echodense material under the TV leaflets (Figure 4C and 4D). Cardiac magnetic resonance imaging was scheduled to delineate the source of TV stenosis. Immune serologies were sent in anticipation of transplantation and were notable for IgG antibodies to Epstein-Barr virus.
Thirty-six days after surgery and 1 day before the scheduled cardiac magnetic resonance imaging, the patient was brought to the emergency department by ambulance after experiencing hemoptysis and collapsing at home. He was asystolic on arrival, and resuscitative efforts were unsuccessful. Autopsy revealed large fungating intracardiac masses in the right ventricle and right atrium with extension onto the prosthetic valve leaflets (Figure 5B). Intraarterial lung masses were present in the distal pulmonary and lobar arteries (Figure 5A) with parenchymal hemorrhage of the bilateral lower lobes. Cardiopulmonary collapse was presumed secondary to rapid tumor recurrence leading to right ventricular outflow tract obstruction and pulmonary embolus with hemorrhage.

IMT is a rare fibroinflammatory spindle cell lesion that most commonly occurs in the lung in childhood or infancy. The clinical course is usually benign, with curative surgical resection or spontaneous regression being the norm. Approximately 40 cases of cardiac IMT have been reported, of which 2 were recurrent. In our patient, IMT first recurred at a different site near the TV annulus 2 years after subtotal resection of a right ventricular septal lesion. It is unclear whether the first recurrence represented progression of the primary tumor given incomplete removal or a metachronous IMT. The long disease-free interval and presence of Epstein-Barr virus antibodies raise the possibility of de novo IMT formation in a predisposed host, perhaps triggered by latent Epstein-Barr virus infection.

The cause of rapid recurrence leading to sudden death shortly after aggressive re-resection is unknown. This case highlights that cardiac IMT can lead to catastrophic outcomes owing to the sensitive location and unpredictable behavior of the tumor. Transplantation may be a consideration for locally aggressive or recurrent cardiac IMT.

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Disclosures
None.

References
Figure 4. Transthoracic echocardiography before and 1 month after resection of the recurrent inflammatory myofibroblastic tumor. A and B, Preoperative images showed a nodular mass covering the posterior wall of the right ventricle and extending anteriorly across the tricuspid valve. The mass appeared to be encasing the anterior and posterior tricuspid valve leaflets, impeding leaflet mobility and coaptation. C and D, Echocardiography 1 month after resection with recurrent tumor in the right atrium and the apex of the right ventricle. A4C indicates apical 4-chamber view; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; nTV, native tricuspid valve; PSAX, parasternal short axis view; pTV, prosthetic tricuspid valve; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract; Pre-op, preoperative; Post-op, postoperative. Arrow denotes the tumor.

Figure 5. Autopsy specimens. A, Left lung with distal left main pulmonary artery tumor embolus and thrombus. B, Right atrial view showing extensive tumor and thrombus infiltration of the prosthetic tricuspid valve (black arrow) and tumor involvement of the right atrial wall (white arrow).
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