A 46-year-old male non-smoker presented to the emergency department with multiple episodes of syncope and resting dyspnea. He reported worsening dyspnea on exertion for the past 4 months. Electrocardiogram showed normal sinus rhythm with non-specific ST-T changes in V1–V4. Echocardiography showed a large mass obliterating most of the right ventricular cavity and extending to the tricuspid leaflets, resulting in severe tricuspid stenosis with turbulent flow across the valve (Figure A, Videos 1, 2). Continuous wave Doppler recording over the tricuspid valve showed mean diastolic gradient of 9.6 mmHg and peak gradient of 18.3 mmHg (Figure B).

Left ventricular function was normal. In view of his symptoms, the patient underwent urgent surgery for resection of the tumor. In surgery, the right ventricle tumor was found to originate from the endocardium of the interventricular septum and extended to the tricuspid valve (Figure C, D). There was chordal involvement. The right ventricular free wall was free of tumor. The tumor was resected and the tricuspid valve was replaced with St. Jude bileaflet prosthesis. Postoperative course was uneventful. Histological examination of the tumor showed polygonal and stellate cells surrounded by abundant loose stroma rich in acid mucopolysaccharides. Direct cardiac muscular invasion and abnormal mitoses were not found. Immunohistochemical staining showed strong immunoreactivity for CD31. Moreover, while the cells over the vascular boundaries were stained with CD34 antigen, they remained negative for calretinin. Typical histologic features indicated myxoma (Figure E-H). At 12-month follow-up, echocardiography showed no evidence of recurrence (Video 3*).