

An unusual cause of severe tricuspid stenosis

Ciddi triküspit darlığının alışılmadık bir nedeni

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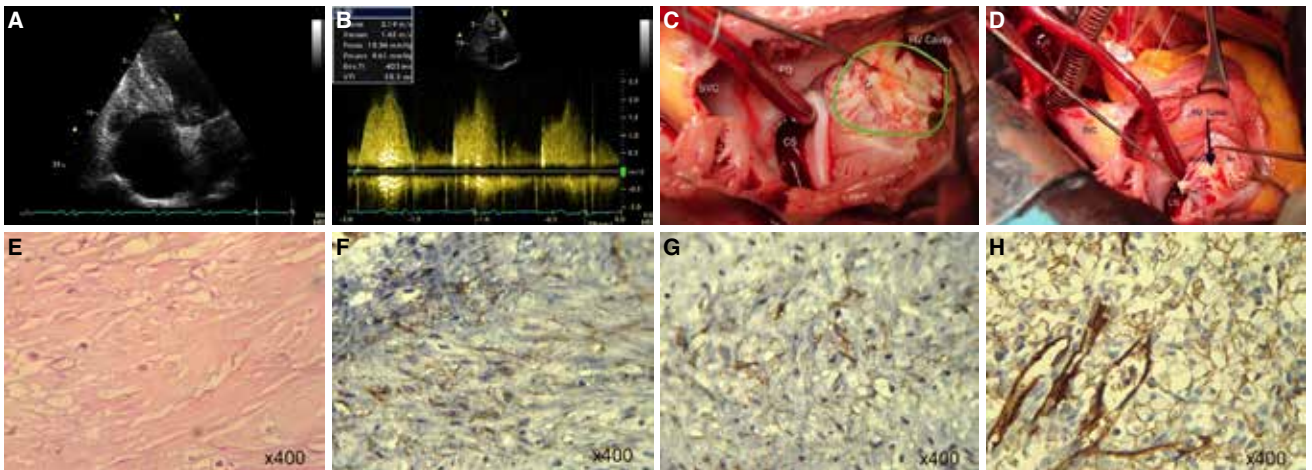
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A 46-year-old male non-smoker presented to the emergency department with multiple episodes of syncope and resting dyspnea. He reported worsening dyspnea on exertion for the past 4 months. Electrocardiogram showed normal sinus rhythm with non-specific ST-T changes in V1–V4. Echocar-

diography 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*).



Figures– (A) Apical 4-chamber echocardiographic image showing obliteration of the right ventricle by the tumor, involvement of tricuspid valve, and right atrial enlargement. IVS: Interventricular septum; LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle. (B) Continuous wave Doppler recording through the tricuspid valve showing mean gradient of 9.61 mmHg and peak gradient of 18.34 mmHg. Surgical view demonstrating near total occlusion of the tricuspid valve (encircled area in C) by the ventricular tumor (arrows in D); the right atrium was draining into the right ventricle through only 2 small orifices. FO: Fossa ovalis; SVC: Superior vena cava; CS: Coronary sinus; RV: Right ventricle; ant: Anterior leaflet; Post: Posterior leaflet. (E) Pathological exam showing round, polygonal, or stellate cells surrounded by abundant loose stroma rich in acid mucopolysaccharides (hematoxylin eosin stain x400). (F) Immunohistochemical staining for calretinin was negative. (G) Immunohistochemical staining showing immunoreactivity for CD34 around vascular boundaries. (H) Immunohistochemical staining showed strong immunoreactivity for CD31. *Supplementary video files associated with this presentation can be found in the online version of the journal.