Images in Cardiovascular Medicine

Large Primary Cardiac Lymphoma

Causing Functional Tricuspid Valve Stenosis

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A previously healthy 49-year-old man presented with an 8-week history of progressive dyspnea on exertion, nonproductive cough, and chills. Physical examination results and laboratory findings suggested right-sided heart failure. A transthoracic echocardiogram (TTE) revealed a large multilobular mass extending from the right atrium through the tricuspid valve (TV) into the right ventricle (Fig. 1). Color-flow Doppler findings at the level of the TV were consistent with functional stenosis (Fig. 2A). The mean gradient across the TV was 9 mmHg at a heart rate of 85 beats/min (Fig. 2B). The uptake of echocardiographic contrast medium enabled us to see the mass further penetrate the myocardial free wall into the pericardium, suggesting neovascularity. Histologic analysis of a biopsy specimen revealed large atypical cells consistent with lymphoma (Fig. 3). A TTE obtained 12 days after chemotherapy began revealed tumor shrinkage (Fig. 4A). The mean gradient had decreased to 3 mmHg, and the patient's symptoms had improved. Four months after chemotherapy had begun, scans obtained with use of positron-emission tomography/computed to-

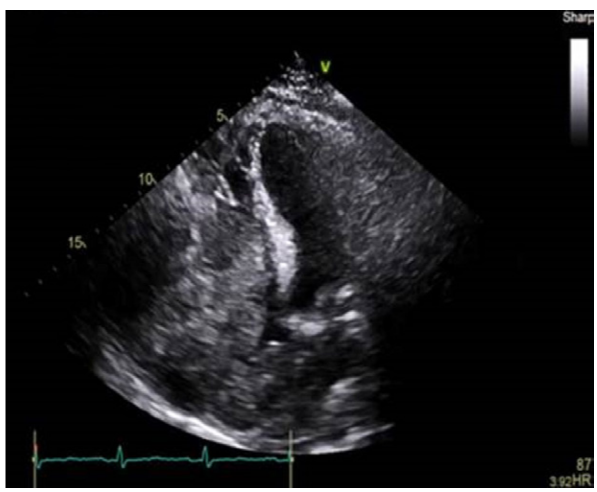

Fig. 1 *Transthoracic echocardiogram shows a large heterogeneous mass occupying the right atrium and right ventricle, with obstruction at the level of the tricuspid valve.*

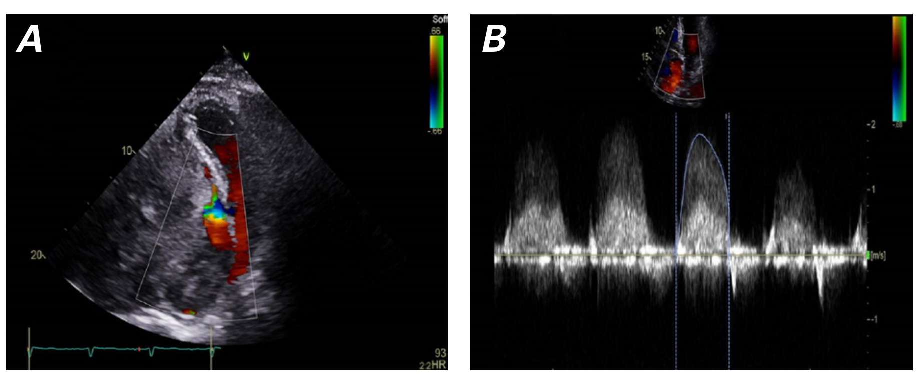

Fig. 2 ***A****) Transthoracic echocardiogram (color-flow Doppler mode) at the level of the tricuspid valve shows functional stenosis.* ***B****) Continuous-flow Doppler mode reveals a peak gradient of 14 mmHg and a mean gradient of 9 mmHg.*

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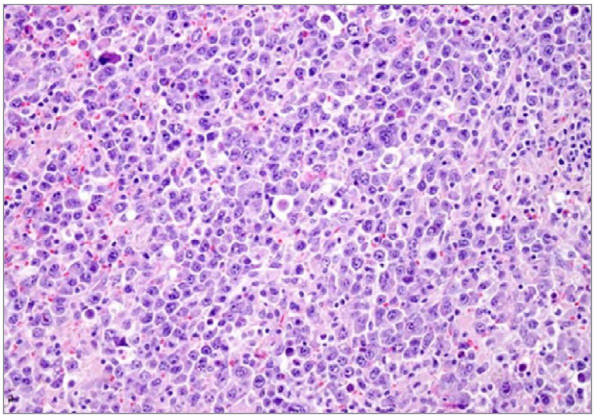

Fig. 3 *Photomicrograph from a biopsy specimen of the mass shows infiltration of cardiac tissue by large atypical cells (H & E, orig. ×100).*

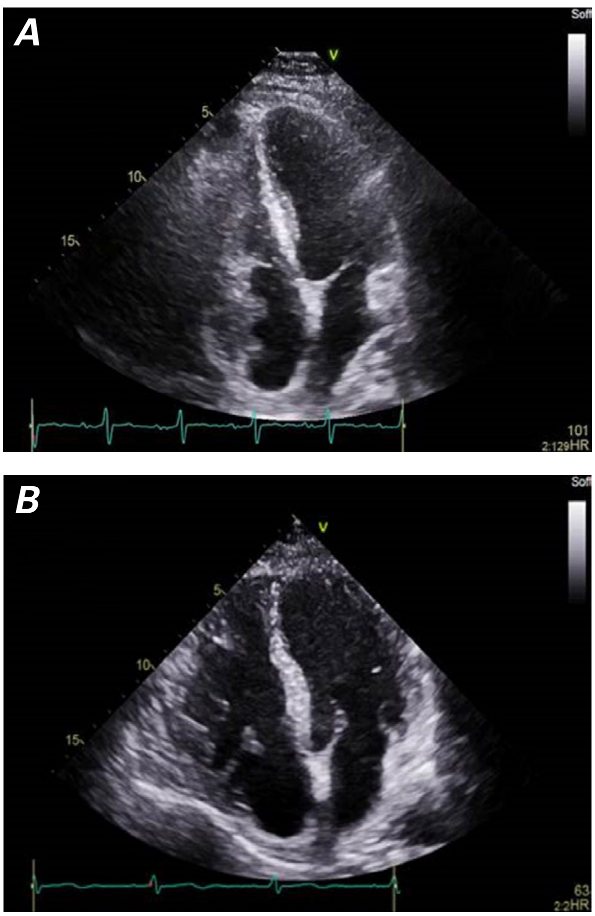

Fig. 4 *Transthoracic echocardiograms.* ***A****) Twelve days after chemotherapy began, the mass was substantially smaller;* ***B****) 18 months later, it had almost disappeared.*

mography showed further tumor shrinkage, from 13.55 × 10.3 to 5.8 × 2.8 cm. Eighteen months after chemotherapy initiation, a TTE showed near-resolution of the tumor (Fig. 4B).

Comment

Primary cardiac lymphoma is a rare malignant tumor that typically affects the right atrium.[1-3] Presenting symptoms include dyspnea, chest pain, arrhythmias, congestive heart failure, and constitutional disorders.[2] Our patient's primary cardiac lymphoma is one of the largest reported in the medical literature.[4]

Early detection is crucial for better outcomes.[5] Rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy is the mainstay of medical therapy.[6] Overall, the response rate to therapy was 84%, and 59% of those tumors resolved.[2]

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