A 13 year-old boy presented with a history of fatigue and abdominal pain. With imaging studies, intra-abdominal and intracardiac tumors were revealed. The tumor almost completely filled the right atrium. It also infiltrated the right ventricular free wall, the interatrial septum, the left atrium, and the pericardium (Figure 1; Video Clip Supplement 1; available at www.jpeds.com). Caval venous and tricuspid inflow was compromised (Figure 2; Video Clip Supplements 2 and 3; available at www.jpeds.com). The patient was placed on intensive care surveillance. Histology, obtained with pericardiocentesis and confirmed with endoscopic gastrointestinal biopsy, revealed a B-cell non-Hodgkin’s lymphoma (B-NHL). Chemotherapy was initiated according to the German National “B-NHL BSM 2004” protocol. The tumor had already disappeared almost completely at 4-week follow-up (Video Clip Supplement 4; available at www.jpeds.com).

Malignant cardiac tumors in children are rare.1-3 They are mostly seen in disseminated B-NHL.2 Not uncommonly, they present in an emergency context with pending hemodynamic compromise.4 Myocardial biopsy has been considered a prerequisite, as has surgery for immediate relief of inflow or outflow obstruction.5 The prognosis has been considered poor, mostly because of a high early and perioperative mortality.1,5 However, data are limited. Management recommendations are confusing. Approaches other than early surgery have been suggested.5 This case adds to the limited literature about such tumors in children. There is growing evidence that detailed assessment can be achieved non-invasively, that histology can reliably be obtained by pericardiocentesis, that sensitivity to chemotherapy of such tumors is usually high, and that surgery can be avoided even in cases of advanced disease.1-3,5 With this less aggressive regimen, prognosis may not be as poor as previously thought.4 More evidence-based treatment recommendations are needed.


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Figure 1. Magnetic resonance imaging, balanced fast-field echo sequence, 4-chamber view. The right atrium is almost completely filled by a hypointense mass (arrowheads). The mass has infiltrated the pericardium, the interatrial septum, and the right ventricular free wall down to the apex. There is pericardial effusion anterior to the right ventricle (asterix).

Figure 2. Magnetic resonance imaging, balanced fast-field echo sequence, parasagittal scan. There is near complete obstruction of the superior vena cava by the tumor (arrow). The infiltrating intracardiac mass (arrowheads) and the pericardial effusion are shown (asterix). MRI, Magnetic resonance imaging; RA, right atrium; LV, left ventricle.
REFERENCES