Cardiac Fibroma in an Infant: Magnetic Resonance Imaging Characteristics

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INTRODUCTION

Primary cardiac neoplasms are uncommon, with a reported prevalence of 0.0017–0.28% of autopsies (1). Approximately 75% of these are benign tumors, of which the most common is myxoma. Cardiac fibroma is a rare benign tumor that primarily occurs in infants and children. Imaging modalities used for diagnosis of cardiac masses include echocardiography, computed tomography, and magnetic resonance imaging (MRI). MRI can depict the anatomic extent of the tumor and the pattern of contrast enhancement. In addition, cine MRI can demonstrate functional pathology of the heart and circulation dynamics. The current report describes an infant with right ventricular fibroma on whom MRI was performed to demonstrate the extent of the tumor and to suggest a specific diagnosis.

CASE REPORT

A 2-month-old boy without significant past medical history presented with upper respiratory infection and 2 days of vomiting. On chest radiographs the mediastinum was markedly widened. Echocardiography revealed a large mass compressing the right atrium and right ventricle. MRI showed that the mass arose from the anterior free wall of the right ventricle and protruded into the anterior mediastinum, displacing the heart posteriorly

Received July 24, 1998; Accepted September 10, 1998 Address reprint requests to C. B. Higgins.

(Fig. 1A) and severely compressing the lumen of the right ventricle, most profoundly in the inflow area (Fig. 1B). The right atrium and the inferior vena cava were dilated secondary to the narrowed inflow region of the right ventricle. Cine MRI demonstrated restriction of right ventricular wall motion in the area of the tumor. A pericardial effusion of moderate size was noted. On electrocardiographically-gated SE T1-weighted images, the mass was homogeneous and isointense to myocardium, and its margins were poorly delineated (Fig. 1, A and B). It contained no areas of signal void, suggesting an absence of calcification. After intravenous administration of gadolinium-diethylenetriaminopentaacetic acid (DTPA) contrast media (0.1 mmol/kg), fatsuppressed SE T1-weighted images demonstrated that the mass was well circumscribed and had irregular peripheral enhancement (Fig. 1C). Because of the enhancement pattern and the circumscribed margins of the mass, a specific diagnosis of fibroma was suggested.

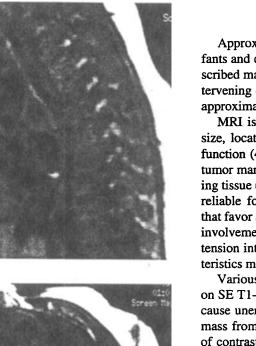
At surgery a firm white mass was removed en bloc from the anterior free wall of the right ventricle. The tumor was connected to the myocardium by a short stalk. Histology demonstrated spindle cells surrounded by collagenous stroma, consistent with a fibroma. No calcification was seen within the tumor.

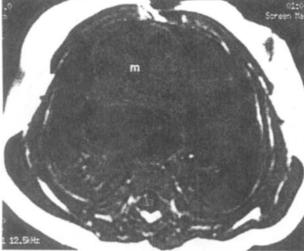
The patient recovered with stable hemodynamic status throughout the postoperative course. One year after surgery, the patient is doing well.

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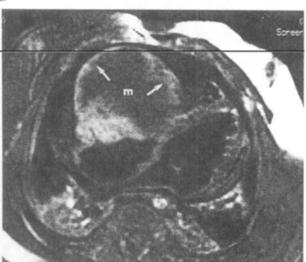
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DISCUSSION

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Approximately 90% of cardiac fibromas occur in infants and children (2). Fibroma is usually a well-circumscribed mass primarily composed of spindle cells and intervening collagen. On histology, calcification is seen in approximately one-half of these tumors (3).

MRI is an effective modality for depiction of tumor size, location, and internal features and altered cardiac function (4,5). Contrast-enhanced MRI can demonstrate tumor margins, extension, and involvement of surrounding tissue (4,5). However, the enhancement pattern is not reliable for determination of malignancy (4). Findings that favor a malignant tumor include pericardial effusion, involvement of more than one cardiac chamber, and extension into the mediastinum (6), although these characteristics may also be associated with benign masses.

Various appearances of fibromas have been described on SE T1-weighted images and cine MRI (4,5,7-9). Because unenhanced MRI cannot distinguish an isointense mass from surrounding myocardium, the administration of contrast media may be necessary to delineate the tumor. In one reported case, heterogeneous contrast enhancement was seen throughout the fibroma, with nonenhancing areas thought to correspond to calcification (7). In two prior cases and in the current case, the cardiac fibroma showed peripheral enhancement (5,8). Because Gd-DTPA is administered intravenously but quickly equilibrates into the extracellular space, this enhancement pattern suggests that the central fibrous tissue is poorly vascularized, whereas the periphery has greater vascularization and a larger extracellular space (8). Because this pattern of enhancement pattern is similar to that of a rapidly growing tumor with central necrosis, it is not reliable to exclude a malignant tumor.

CONCLUSION

In patients with a cardiac mass, MRI can be used define the precise location and extent of the tumor. In the current case, MRI indicated that the compression of the

Figure 1. Right ventricular fibroma: electrocardiogram-gated SE T1-weighted MRI. (A) Sagittal image (TR 365, TE 20) shows the mass (m) displacing the heart posteriorly. (B) Axial image (TR 370, TE 20) shows a homogeneous mass (m) of intermediate signal intensity (similar to normal myocardium) arising from the right ventricular wall. (C) Axial fat-suppressed image (TR 379, TE 20) after intravenous administration of Gd-DTPA shows the circumscribed mass (m) with complete irregular rim enhancement (arrows). The center of the tumor demonstrates signal intensity similar to that of myocardium.

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right ventricular cavity was caused by a mural mass rather than an intracavitary mass.

ACKNOWLEDGMENT

Supported in part by a research training grant from the National Institutes of Health (T32-HL07570) (to G.P.R.).

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