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Congenital & Pediatric: Case Report

Epicardial Involvement of Constrictive Pericarditis Can Be Detected by Magnetic Resonance Imaging

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Constrictive pericarditis is a surgical disease that requires removal of the pericardium. In cases in which the disease process involves the epicardium, removing the pericardium may not adequately treat the constrictive process. Current imaging techniques are limited in their ability to preoperatively determine epicardial involvement. Cardiac magnetic resonance imaging with late gadolinium enhancement of the pericardium is useful in conjunction with echocardiography to define extent of disease, pericardial compliance, and hemodynamics. We propose the use of cardiac magnetic resonance imaging, and specifically late gadolinium enhancement extending into the epicardium, as a marker of epicardial involvement based on our initial experience.

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We present 2 cases of constrictive pericarditis in which cardiac magnetic resonance imaging (MRI) with late gadolinium enhancement (LGE) helped define the extent of pericardial and epicardial involvement. Both patients required epicardiectomy in addition to pericardiectomy to relieve the constriction.

CASE REPORTS

PATIENT 1. A 17-year-old girl with Myhre syndrome and tetralogy of Fallot, for which she underwent complete repair at 2 months of age with transannular patch and ventricular septal defect closure, presented with shortness of breath.

Cardiac MRI demonstrated significant pericardial thickening (up to 7 mm) and LGE of this thickened pericardium that extended to the epicardium but otherwise spared the myocardium. There was no pericardial thickening or delayed enhancement along the anterior surface of the heart where the pericardium had been removed at the time of tetralogy repair. There was also a large right-sided pleural effusion (Figure 1). During cardiac catheterization, 1 L of serous fluid was evacuated on chest tube placement. Cardiac catheterization yielded the following pressures: mean right atrium, 23 mm Hg; right ventricle, 57/24 mm Hg; pulmonary artery, 49/23 mm Hg; pulmonary capillary wedge pressure, 23 mm Hg; and left ventricle, 91/25 mm Hg. Echocardiography demonstrated normal systolic biventricular function, severe pulmonary insufficiency, and no other valvular or intracardiac abnormalities. She was referred for surgical management of her presumed constrictive pericarditis.

An off-pump subtotal pericardiectomy was performed, leaving only bilateral pedicles around each phrenic nerve. After pericardiectomy, there was still evidence of epicardial thickening, so further epicardiectomy was performed. Subsequent spot measurement of left atrial pressure was 8 mm Hg. The right atrial pressure measured 10 mm Hg. Intraoperative echocardiography demonstrated normalization of mitral inflow patterns, without evidence of respiratory variation, and normal biventricular function.

The patient recovered well from surgery but experienced prolonged chest tube drainage. She was discharged on postoperative day 14. Gross examination of the excised pericardium and epicardium demonstrated thickened fibromembranous tissue. Microscopic examination of both specimens confirmed fibrosis with patchy areas of acute and chronic inflammation with hemosiderin deposition (Figure 1).

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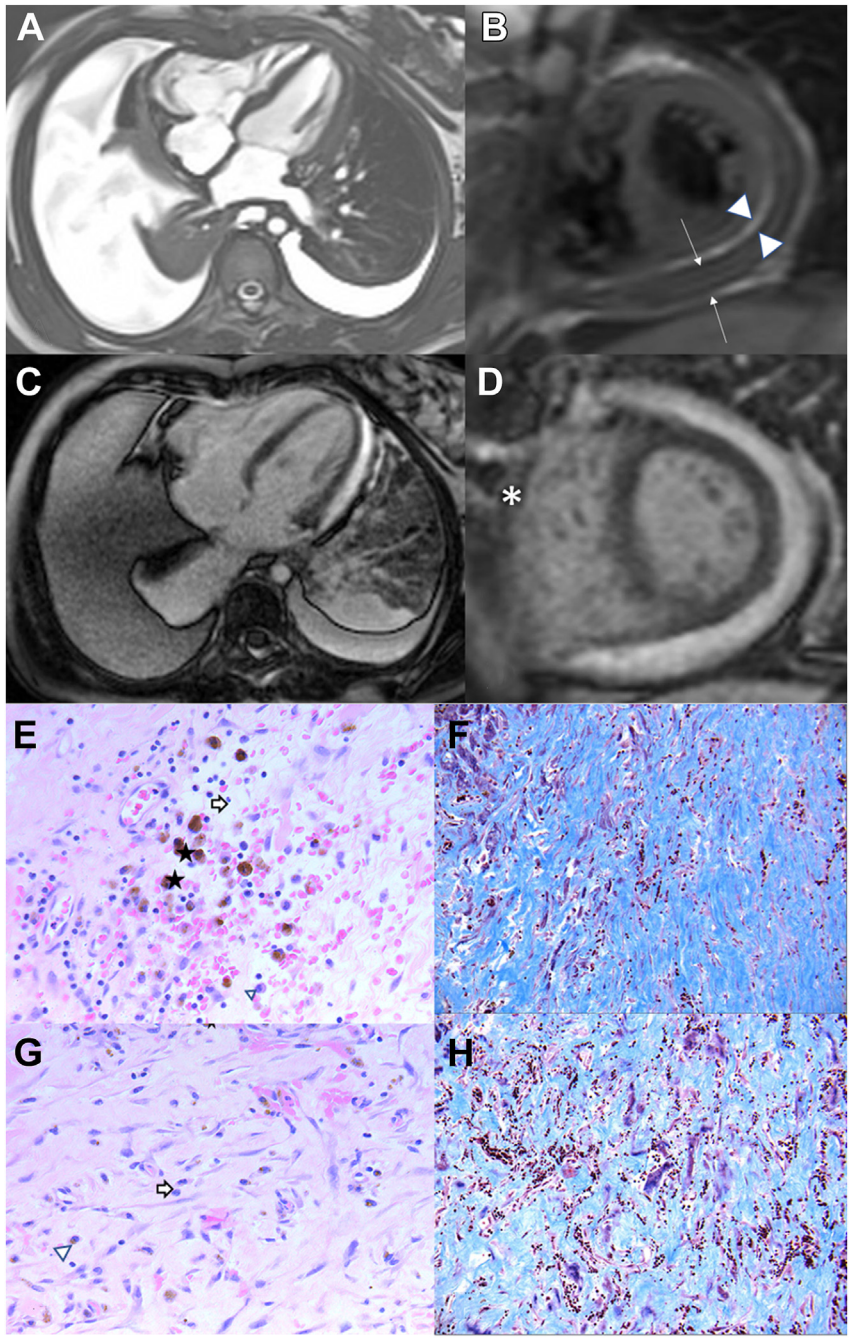

FIGURE 1 Cardiac magnetic resonance imaging and histopathologic evaluation of patient 1. (A) Steady-state free precession image acquired in the 4-chamber view demonstrates a large right pleural effusion and a smaller left pleural effusion. The left ventricle lateral wall appears thickened, but the pericardium is not distinguishable from myocardium. (B) T1-weighted dark blood image acquired in the short-axis view highlights 7-mm pericardial thickening (arrows) and bright signal in the epicardial region (arrowheads). (C, D) Inversion recovery delayed enhancement image acquired in 4-chamber and short-axis views demonstrates 9-mm pericardial late gadolinium enhancement that encroaches on the epicardium especially along the inferior-lateral wall of the left ventricle. Delayed enhancement in (D) confirms that the epicardial bright signal observed in (C) is also composed of fibrosis. There is no anterior pericardial thickening or late gadolinium enhancement (*) where the pericardium was removed at the time of tetralogy repair. (E, F) Epicardial and (G, H) pericardial histopathology sections are shown. (E, G) Hematoxylin and eosin–stained sections at magnification ×400 show acute and chronic inflammation composed of neutrophils (arrows) and macrophages (arrowheads) with foci of hemosiderin deposition (stars). (F, H) Masson trichrome stain shown at magnification ×200 highlights collagen deposition in areas of fibrosis.

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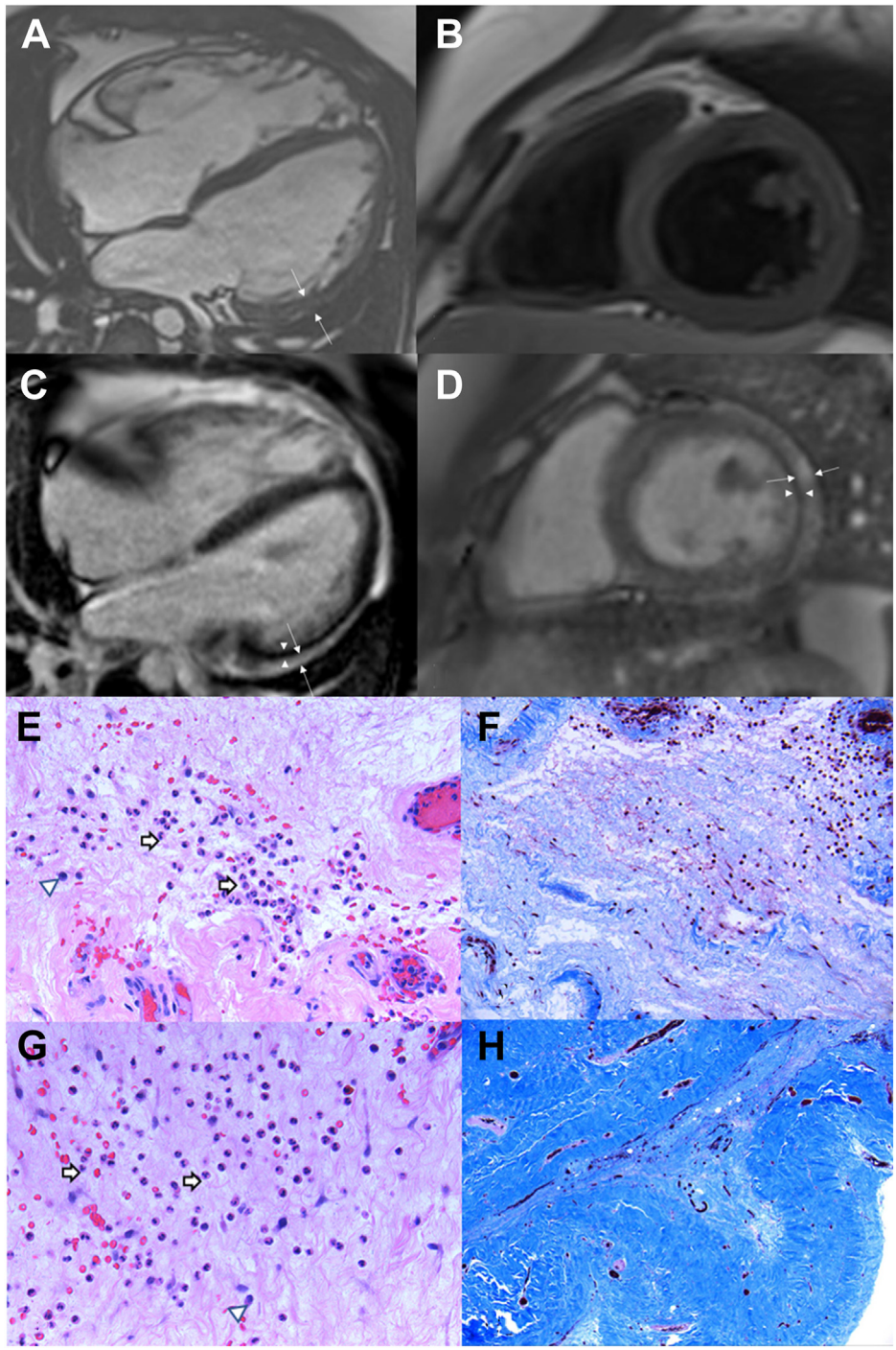

FIGURE 2 Cardiac magnetic resonance imaging and histopathologic evaluation of patient 2. (A) Steady-state free precession image acquired in the 4-chamber view demonstrates that there is no pericardial fluid and normal left ventricle wall thickening. The left ventricle lateral wall measures up to 7 mm (arrows). There is pericardial fat along the left ventricle apex. (B) T1-weighted dark blood image acquired in the short-axis view demonstrates that there is no definitive pericardial thickening. (C, D) Inversion recovery delayed enhancement image acquired in 4-chamber and short-axis views demonstrates significant pericardial late gadolinium enhancement along the lateral wall of the left ventricle that extends into the epicardium (arrows). The adjacent region of normal (dark) myocardium without late gadolinium enhancement represents the subendocardium and midmyocardium (arrowheads). (E, F) Epicardial and (G, H) pericardial histopathology sections are shown. (E, G) Hematoxylin and eosin–stained sections at magnification ×400 show acute and chronic inflammation composed of neutrophils (arrows) and macrophages (arrowheads). (F, H) Masson trichrome stain shown at magnification ×200 highlights collagen deposition in areas of fibrosis.

PATIENT 2. A 16-year-old boy with a history of pericardial effusion presented with emesis and orthopnea. Two and a half years earlier, computed tomography angiography showed pericardial thickening, effusion, and reactive lymph nodes. Infectious workup was unrevealing, and he was treated with anti-inflammatory medications.

His symptoms persisted, and he was ultimately diagnosed with idiopathic inflammatory pericarditis and transitioned to the immunomodulator canakinumab. However, the patient continued to experience exertional dyspnea and tachycardia.

Cardiac catheterization revealed the following pressures: mean right atrium, 12 mm Hg; right ventricle, 32/14 mm Hg; pulmonary artery, 30/18 mm Hg; pulmonary capillary wedge pressure, 15 mm Hg; and left ventricle, 91/16 mm Hg. The presence of elevated left- and right-sided filling pressures was concerning for diastolic dysfunction and constrictive physiology. Cardiac MRI T1-weighted imaging demonstrated borderline pericardial thickening (3 mm) without evidence of myocardial edema. There was significant LGE of the pericardium that extended into the epicardium (Figure 2).

These data were consistent with constrictive pericarditis, and the patient was referred for pericardiectomy. An off-pump subtotal pericardiectomy was performed, sparing the phrenic nerves as pedicles of pericardium. The surface of the heart was edematous and covered by a gel-like thickened epicardium, which was also resected. After the procedure, right and left atrial pressures were 8 to 9 mm Hg with improving symptoms.

The patient's postoperative course was unremarkable, and he was discharged on postoperative day 3. Epicardial and pericardial tissue demonstrated similar findings grossly, and tissue sections were seen to show variably dense fibroconnective and adipose tissue with patchy acute and chronic inflammation on histologic review (Figure 2).

COMMENT

Advanced imaging techniques, including cardiac MRI with LGE, are important adjuncts in the workup of constrictive pericarditis.[1-7] Here we present 2 cases in which LGE of the pericardium and encroachment on the myocardium informed operative planning and the decision to pursue epicardiectomy in addition to pericardiectomy.

In patient 1, LGE on cardiac MRI confirmed pericardial enhancement and extension of the LGE into the subepicardium, suggesting that the inflammation impinges on the subepicardium. Histologic evaluation showed little demarcation between the epicardium and pericardium because of the presence of dense collagen deposition, fibrosis, and tissue remodeling that correlated well with the imaging findings. In patient 2, pericardial thickening was borderline on cardiac MRI based on T1-weighted imaging, but there was significant LGE extending beyond the pericardium. These findings correlated with the histopathologic evidence of similar lymphocytic infiltration and collagen deposition in both the pericardium and epicardium.

These 2 cases illustrate the novel application of LGE to preoperatively assess the extent of epicardial involvement in cases of constrictive pericarditis. This is a preliminary report of a phenomenon that was determined retrospectively. Prospective assessments, larger patient numbers, and further validation will be necessary to determine the utility of these measurements in surgical planning for pericardiectomy in patients with constrictive pericarditis.

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DISCLOSURES

The authors have no conflicts of interest to disclose.

PATIENT CONSENT

Informed consent was obtained from the legal guardians of both patients.

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