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CONGENITAL HEART DISEASE

Cardiovascular magnetic resonance in endocardial fibroelastosis

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Endomyocardial fibroelastosis is a rare disorder marked by characteristic morphological changes of the endocardium. We report a case of endomyocardial fibroelastosis diagnosed by late gadolinium-enhanced CMR and confirmed by endomyocardial biopsy. The patient improved following treatment with imatinib.

Key Words: Endomyocardial fibroelastosis; Eosinophilia; Cardiovascular magnetic resonance imaging; Endomyocardial biopsy

1. Introduction

Endomyocardial fibroelastosis (EFE) presents in a primary form that manifests in infancy as well as secondary forms related to hypereosinophilia and leukemic disorders (1). Accurate identification has previously required endomyocardial biopsy, though cardiovascular magnetic resonance (CMR) is uniquely suited to visualize endomyocardial fibrosis noninvasively. We present a case demonstrating CMR findings in EFE and recommendations for its role in diagnosis of this disease.

2. Case report

A 23-year-old African-American male presented to the emergency department with 1 month of progressive fatigue and acute visual changes. Physical examination was notable for tachycardia, pale conjunctiva, and ophthalmologic findings suggestive of retinal emboli. Complete blood count demonstrated profound eosinophilia, with an absolute eosinophil count of 22,000/μL. Electrocardiography demonstrated diffuse T-wave inversion. Echocardiography revealed bright echodensity along much of the LV endocardium and visualized portions of the RV endocardial surface.

CMR was performed on a 1.5 Tesla scanner (CV*i*, GE Healthcare) using breath hold, electrocardiographically gated pulse sequences. Intravenous gadolinium-DTPA (0.2 mmol/kg) was administered and late gadolinium myocardial enhancement images were obtained in multiple planes after 10 minutes using standard techniques (2). Multiple acquisitions using an inversion recovery-prepped gradient echo sequence were obtained with incrementally varied inversion times (TIs) until optimal contrast between normal and abnormal myocardium was obtained with TI = 250 ms. There was extensive endomyocardial enhancement involving the mid and apical left ventricle as well as the right ventricle (Fig. 1). Mural thrombus was also identified. The diagnosis of EFE was confirmed by microscopic examination of endomyocardial biopsy tissue (Fig. 2).

After a lack of response to corticosteroids, the patient was started on imatinib mesylate with immediate decline in absolute eosinophil count and clinical improvement. Treatment also included initiation of beta-blockade and angiotensin-converting enzyme inhibition for LV dysfunction, as well as anticoagulation for intracardiac thrombus.

3. Discussion

Endocardial fibroelastosis (EFE) is a disease process characterized pathologically by diffuse thickening of the endocardium with layering of collagen and elastic fibers (3). Most commonly described in pediatric populations, there is much controversy as to the existence of EFE as a primary disease process (4). Case reports of EFE in adults most often describe concomitant disorders including mucopolysaccharidoses, valvular heart disease, and congenital heart disease. Common

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clinical presentations include left ventricular dysfunction, congestive heart failure, and sudden cardiac death (5). Conventional diagnostic modalities may provide nonspecific findings such as left ventricular hypertrophy by electrocardiography. Echocardiography has been the mainstay for diagnosis, particularly in the neonatal and pediatric population (1). Definitive diagnosis has historically required endomyocardial biopsy to identify the characteristic endocardial fibrosis that is often preceded by an inflammatory stage (6).

Computed tomography (CT) allows excellent visualization of cardiovascular calcification. Wang and colleagues described five cases of EFE diagnosed with electron beam CT, noting apical calcification in all (7). They also inferred detection of fibrosis with cardiac CT, though this approach has not been as extensively validated with histopathology as it has with late gadolinium enhancement CMR. Furthermore, assessment of cardiovascular physiology, an essential complement to anatomical detail in selecting therapies and determining prognosis, remains cumbersome with cardiac CT.

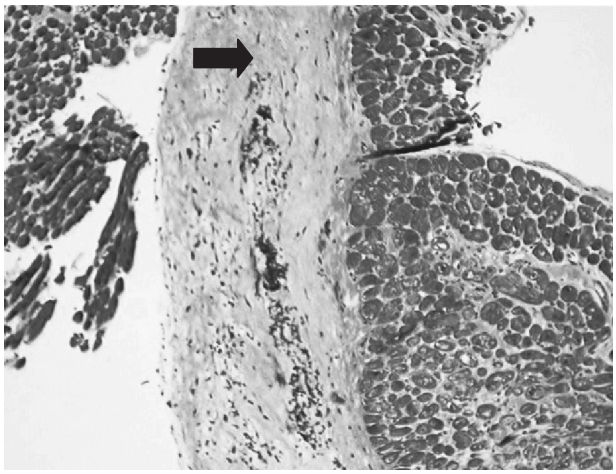
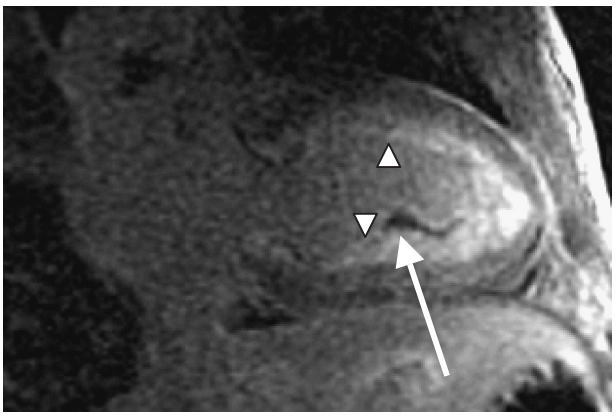


Figure 2. Endomyocardial biopsy, trichrome stain. Arrow denotes area of thickened, fibrotic endocardium.

a



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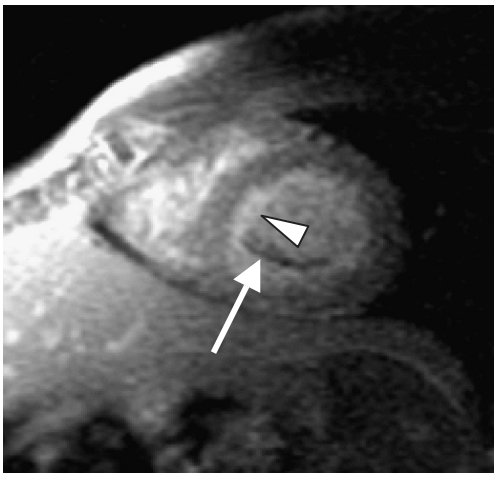

Figure 1. a. Horizontal long-axis (two-chamber) late gadolinium enhancement image demonstrating subendocardial fibrosis (arrowheads) and mural thrombus (straight arrow) of left ventricle. b. Short-axis late post-gadolinium image demonstrating biventricular involvement of fibrosis (arrowhead) and redemonstration of left ventricular mural thrombus (straight arrow).

In ischemic as well as nonischemic cardiomyopathies, late gadolinium enhancement CMR has become the preferred modality for noninvasive detection and quantification of myocardial fibrosis due to its superior spatial resolution as well as its comprehensive capabilities for cardiac anatomical and physiological assessment. This technique currently requires careful optimization of the inversion time to obtain adequate contrast between enhanced and normal myocardium (2). MR can detect endocardial fibrosis and thickening as well as quantify regional wall motion, visualize mural thrombus formation, and quantify left and right ventricular function, all of which are relevant to diagnosis and prognosis for patients with EFE. Additionally, serial evaluation may provide noninvasive assessment of disease progression and treatment efficacy, particularly as novel antifibrotic therapies are implemented. Thus, we recommend that CMR be evaluated further as the diagnostic modality of choice in this disorder.

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