Surgical management of adult endocardial fibroelastosis

Joshua L. Chan, MD,^a Douglas R. Rosing, MD,^b Amy D. Klion, MD,^c and Keith A. Horvath, MD,^a
Bethesda, Md

From the ^aCardiothoracic Surgery Research Program, and ^bCardiovascular and Pulmonary Branch, National Heart, Lung, and Blood Institute, and ^cLaboratory of Parasitic Diseases, National Institute of Allergy and Infectious Diseases, National Institutes of Health, Bethesda, Md.

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Address for reprints: Keith A. Horvath, MD, Building 10-CRC, Room B2-3701, 10 Center Dr, Bethesda, MD 20892 (E-mail: khorvath@aamc.org).

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Histologic demonstration of extensive endocardial fibroelastosis with Russell-Movat staining.

Central Message

Adult-onset endocardial fibroelastosis is a rare condition that in severe cases can precipitate a restrictive cardiomyopathy requiring decortication.

See Editorial Commentary page e85.

▶ Video clip is available online.

CLINICAL SUMMARY

A 37-year-old Caucasian man with inflammatory bowel disease receiving chronic mesalamine therapy presented for evaluation of progressive shortness of breath, exercise intolerance, and fatigue. Physical examination was notable for a grade III/VI holosystolic murmur at the apex radiating to the axilla. Complete blood count results were unremarkable. Electrocardiography revealed an incomplete left bundle branch block with lateral repolarization abnormalities and poor anterior R wave progression. Transthoracic echocardiography and cardiac magnetic resonance (CMR) imaging identified increased echodensity along the inferolateral and inferior walls, extending to the apex. The lesion was associated with hypokinesis of the adjacent wall that involved the subvalvular apparatus. This contributed to abnormal ventricular relaxation and severe mitral regurgitation. CMR also demonstrated an extensive, nonischemic pattern of endocardial delayed enhancement (Figure 1). These findings raised concerns that an abnormal endocardial fibrotic process was restricting ventricular compliance.

Despite an aggressive course of atorvastatin, lisinopril, furosemide, and beta-blockers for treatment of congestive cardiomyopathy, the patient became progressively symptomatic and ultimately required surgical resection of the endocardial thickening (Video 1). Through a transmitral approach, a calcified fibrotic left ventricular mass with near obliteration of the apex was identified, with extension toward the subvalvular apparatus resulting in chordal

retraction. Sharp dissection of the grossly thickened endocardium was accomplished with the aid of 4-0 Prolene retraction sutures, revealing a clearly demarcated and compliant myocardium without residual fibrosis (Figure 2). Mitral valve repair to address the significant annular dilation was performed with placement of a mitral annuloplasty band.

Postprocedure transesophageal echocardiography confirmed improved ventricular compliance, with an E/A ratio of 1.4 compared with 2.6 preoperatively and a deceleration time of 217 ms compared with 92 ms preoperatively. In addition, the mitral valve appeared to be functioning well without significant regurgitation. Histopathologic evaluation with multiple stains (Figure 3), including Russell-Movat pentachrome stain, revealed proliferation of dense collagen and elastin fibers, confirming the diagnosis of endocardial fibroelastosis. Eosinophil peroxidase (EPX) immunohistochemical staining demonstrated rare eosinophils with patchy perivascular eosinophil granule protein deposition.

DISCUSSION

Endocardial fibroelastosis was recognized as early as 1901 by Josserand and Gallavardin, who described a primary myocardial disease following subacute myocarditis. A similar pathological picture was described by Löffler 35 years later in an entity associated with

FIGURE 1. Cardiac magnetic resonance imaging with 3-chamber view. Hypointensity of the endocardium with apical filling defect apparent during steady-state free precession sequences (A), followed by diffuse enhancement with central focus of thrombus during late gadolinium enhancement (B). (Image courtesy of Sujata M. Shanbhag, MD, National Heart, Lung, and Blood Institute.)

eosinophilia, although the term “endocardial fibroelastosis” was first introduced by Weinberg and Himelfarb in 1943.¹ This case highlights a rare and unusual instance of extensive endocardial fibroelastosis in an adult that resulted in severe restrictive cardiomyopathy.

The diagnosis of endocardial fibroelastosis can be challenging owing to its nonspecific clinical presentation. Ejection fraction may be preserved despite clinical evidence of heart failure due to impairment of both systolic and diastolic function. CMR is useful in establishing the diagnosis before obtaining endomyocardial biopsy; characteristic findings include a hypointense endocardial layer during perfusion phase, followed by a hyperintense signal during myocardial delayed-enhancement sequences.² If a biopsy is performed, specific fibroelastic staining can provide additional confirmation. Histologic evidence of extensive abnormal endocardial deposition of

elastin and collagen is characteristic, with minimal penetration into the underlying myocardium.

Medical treatment, including diuretics, is typically directed at the management of heart failure. Although reports describing the surgical approach to endocardial fibroelastosis are limited, operative indications include cases refractory to pharmacologic therapy and progressive mechanical impairment of ventricular function.³ Diffuse endocardial thickening affecting predominantly the left ventricle is frequently observed, and is addressed with sharp decortication until removal of residual fibrosis is achieved.

FIGURE 2. Decortication of the left ventricular fibrotic process through a transmitral approach. With the aid of a pledgeted 4-0 Prolene suture, the lesion was sharply dissected off the myocardium.

Surgical Management of Adult Endocardial Fibroelastosis

VIDEO 1. Operative steps describing the surgical management of adult endocardial fibroelastosis. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(17\)31095-4/addons](http://www.jtcvsonline.org/article/S0022-5223(17)31095-4/addons).

FIGURE 3. Histology of endocardial biopsy at the time of surgery. A, Hematoxylin and eosin staining showing dense fibrous tissue (4 $\times$) and focal areas of chronic lymphocytic inflammation with rare eosinophils (*inset*, 20 $\times$). B, Russell–Movat pentachrome stain showing dense proliferation of collagen and elastin fibers. C and D, Eosinophil peroxidase immunohistochemical staining demonstrating granule protein deposition around isolated vessels (40 $\times$) (C) and in areas of inflammation (40 $\times$) (D).

Adequate resection is confirmed with exposure of normal-appearing compliant myocardium; improved ventricular mechanics can be verified by echocardiography as well.

Believed to be a reactive and proliferative process in response to myocardial stress, hypereosinophilia is the most frequent etiology of endocardial fibroelastosis, although other inciting factors, such as myocarditis and autoimmune diseases, have been implicated. In this specific case, all usual etiologies were excluded. Thorough evaluation for hypereosinophilic syndrome was negative, including *FIP1L1/PDFGRA* and *JAK2* mutations. Although immunohistochemical staining revealed patchy perivascular deposition of EPX, this is likely a reflection of tissue remodeling and has been described in the setting of noneosinophilic cardiomyopathy.⁴ Metastatic workup, nutritional evaluation, and viral serology tests were not revealing; however, on examination of possible drug-related etiologies, the administration of mesalamine (5-aminosalicylic acid) was suspected as a potential trigger for myocarditis-induced endocardial fibroelastosis. This generally well-tolerated treatment for inflammatory bowel disease has been associated with myocarditis in rare instances secondary to drug hypersensitivity.⁵ Although its contribution was speculative, mesalamine was discontinued to potentially prevent

additional episodes of myocarditis-induced endocardial fibroelastosis.

CONCLUSIONS

Endocardial fibroelastosis is a proliferative disorder that in rare instances can lead to heart failure in adults. In severe cases, surgical management with left ventricular decortication is warranted.

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