Caseous calcification of the anterior mitral valve annulus presenting as intracardiac mass

Sotiris C. Stamou, MD, Alan C. Braverman, MD, and Nicholas T. Kouchoukos, MD, St Louis, Mo

Caseous calcification of the mitral annulus occurs in approximately 0.5% to 1% of patients with mitral annular calcification and most commonly involves the posterior annulus.¹ It can cause mitral regurgitation, mitral stenosis, and heart block, and increases the complexity of mitral valve replacement.² We present a case of caseous calcification of the anterior leaflet of the mitral valve presenting with a retinal artery embolization, severe mitral regurgitation, and a cardiac mass.

CLINICAL SUMMARY

An 84-year-old man presented with loss of vision in the right eye and exertional dyspnea. Fundoscopic examination was consistent with retinal artery embolism. A subsequent evaluation that included a transesophageal echocardiogram (Figure 1) demonstrated a large mass beneath the anterior leaflet of the mitral valve. The echocardiogram also showed severe mitral regurgitation, mild mitral stenosis, and calcified aortic valve leaflets. Computed tomography of the chest showed a calcified mass in the region of the anteroseptal mitral annulus that measured 4.3×3.0 cm, abutting on the atrial aspect of the anterior leaflet of the mitral valve (Figure 2). Operation was recommended.

After establishing cardiopulmonary bypass and cardioplegia, a left atriotomy was performed. Inspection of the mitral valve did not demonstrate a discrete mass. However, caseous material was exuding from the ventricular surface of the anterior leaflet of the mitral valve. An oblique aortotomy was made and extended into the noncoronary sinus. The calcific aortic leaflets were excised, and the thickened and fluctuant anterior leaflet of the mitral valve was incised. Approximately 10 mL of a creamy white liquid was removed. The anterior leaflet was excised through the left atrium. The aortomitral curtain was reconstructed through the aortotomy using a bovine pericardial patch (Figure 3). The mitral and aortic valves were replaced using bioprosthetic valves. A postoperative transesophageal echocardiogram showed

FIGURE 1. Transesophageal echocardiography showing the mass under the anterior leaflet of the mitral valve (arrow). LA, Left atrium; LV, left ventricle.

obliteration of the cavity and no paravalvular regurgitation (Figure 4).

DISCUSSION

Caseous calcification or *liquefaction necrosis* of the mitral valve annulus is a rare variant of mitral annular calcification and typically involves the posterior annulus.^{1,3} In a series of 19 patients with this condition reported by Harpaz and colleagues,¹ isolated involvement of the anterior leaflet

FIGURE 2. Computed tomography scan showing a calcified mass in the region of the anterior mitral valve.

From the Division of Thoracic and Cardiovascular Surgery, Missouri Baptist Medical Center, St Louis, Mo.

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Address for reprints: Sotiris C. Stamou, MD, 3009 N Ballas Road, 1315 Whispering Pines Dr, Suite 266C, Apt L, St Louis, MO 63131 (E-mail: cvsisfun@hotmail.com). J Thorac Cardiovasc Surg 2010;140:e9-10

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FIGURE 3. Postoperative transesophageal echocardiography showing obliteration of the cavity.

was not observed and only 1 patient had involvement of both the anterior and posterior annulus. The prevalence in necropsy series has been reported to be 2.7%.¹ Caseous calcification may present as intracardiac mass or cavity. The contents of the cavity are composed of a mixture of calcium, fatty acid, and cholesterol with a “toothpaste-like” texture.

Most patients with this condition are asymptomatic, and the finding of caseous calcification is an incidental echocardiographic finding. Symptomatic patients have evidence of mitral stenosis or regurgitation, resulting from restricted movement of the mitral leaflets.¹ Of interest, 5 of the 19 patients (23%) reported by Harpaz and colleagues¹ had a history of cerebrovascular events. The rarity of caseous calcification of the mitral valve may lead to misdiagnosis of this entity as an abscess or cardiac tumor. Typical echocardiographic features include a round echo-dense mass with smooth borders and a central area of echolucency resembling liquefaction.^{1,3}

CONCLUSIONS

Asymptomatic patients with caseous calcification of the mitral valve can be managed conservatively. Embolization and symptoms of mitral valve stenosis or regurgitation are indications for surgical intervention. This requires removal of the contents of the cavity and mitral valve replacement. Mitral valve repair is not advisable because unroofing of

FIGURE 4. Intraoperative view through the aortic root. A pericardial patch is used to close the cavity and reconstruct the aortomitral curtain. Ao, Aorta; LA, left atrium; LV, left ventricle; MVP, mitral valve prosthesis.

the cavity will allow necrotic calcific debris to escape into the bloodstream.² Insertion of a bioprosthetic mitral valve and positioning the valve struts over the cavity to allow compression of the cavity may be beneficial.² In our patient the caseous calcification was present within the anterior leaflet of the mitral valve, and reconstruction of the aortomitral curtain with a bovine pericardial patch was required to exclude the cavity from the bloodstream and to provide suitable tissue to anchor both the aortic and mitral valve prostheses. To our knowledge, caseous calcification of the anterior mitral annulus requiring surgical intervention has been reported in only 1 patient who had involvement of the posterior annulus as well and underwent mitral valve replacement.¹ Thus, our patient represents the first reported case of isolated anterior annular calcification treated surgically.

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