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CASE REPORTS

Intracardiac pseudotumor caused by mitral annular calcification

E.A. de Vrey^{a,*}, A.J.H.A. Scholte^a, X.H. Krauss^a, R.A. Dion^b,
D. Poldermans^c, E.E. van der Wall^a, J.J. Bax^a

^a Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands

^b Department of Thoracic Surgery, Leiden University Medical Center, Leiden, The Netherlands

^c Department of Cardiology, Erasmus Medical Center, Rotterdam, The Netherlands

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Abstract The current report describes a rare case of a pseudotumor in the left ventricle. Transthoracic and transesophageal echocardiography demonstrated a round, echodense, mobile mass attached to the posterior mitral leaflet and annulus. At surgical exploration caseous annular calcification of the posterior mitral leaflet was diagnosed. After resection of the mass, successful mitral valvular plasty was performed. Review of the literature indicated that mitral annular calcification is associated with an increased risk of stroke. Optimal treatment may be surgery, especially when valve plasty can be performed, although randomized trials are currently lacking.

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Case report

A 61-year-old man was admitted to the hospital for further evaluation of an intracardiac mass detected at echocardiography in a referring hospital. The medical history consisted of hypertension, hypercholesterolemia, obesity and diabetes mellitus type 2 with secondary renal dysfunction. At presentation, he complained of mild shortness of breath at exercise and claudication. At physical examination,

no abnormalities were observed apart from obesity. Laboratory measurements were normal except elevated serum creatinine (288 mmol/l).

Transthoracic echocardiography was performed (Fig. 1) which showed a round, echodense mass in the left ventricle, attached to the posterior mitral leaflet. There was minimal mitral regurgitation without stenosis. There was left ventricular hypertrophy with normal LV dimensions (LVEDD 55 mm, LVESD 35 mm) and normal function (FS 38%, biplane LVEF 58%). Transesophageal echocardiography demonstrated a round, lobulated, inhomogeneous echodense, partly mobile, mass of 2 × 3 cm,

* Corresponding author. Tel.: +31 71 5283944.

E-mail address: e.a.de_vrey@lumc.nl (E.A. de Vrey).

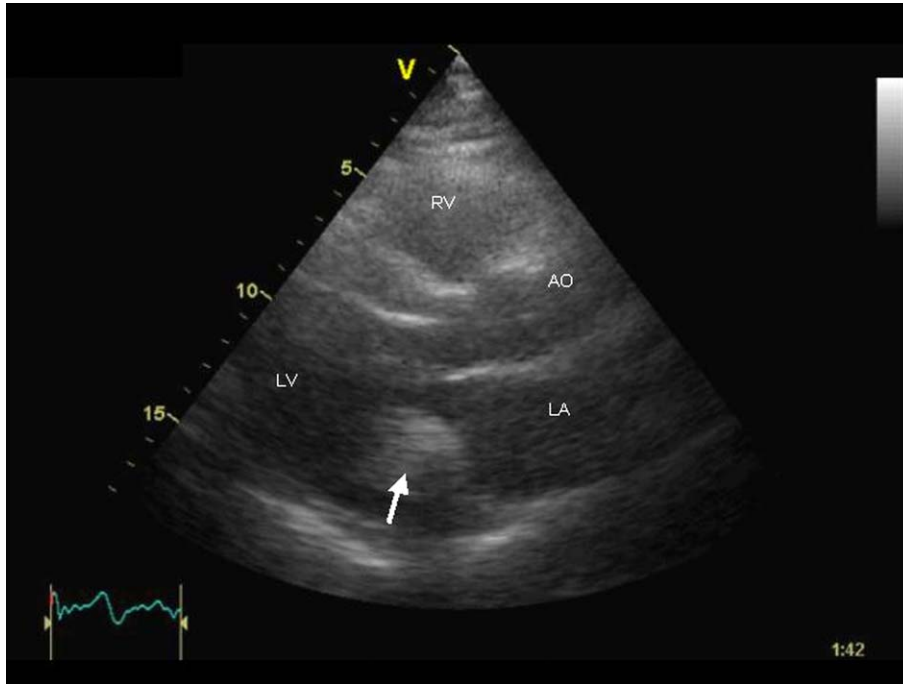


Figure 1 Transthoracic long-axis view demonstrating a tumor-like structure, 2×3 cm, attached to the posterior mitral leaflet of annulus (arrow). LA, left atrium; LV, left ventricle; RV, right ventricle; Ao, aorta.

attached to the posterior mitral annulus (Fig. 2). Furthermore, protrusion of the tumor into the left atrium was observed. The subvalvular mitral apparatus was intact. Left atrial appendage showed no thrombus.

Based on the size, mobility and location of the mass, the risk of embolization in this patient was estimated high. The tumor was considered to be either a myxoma or caseous mitral annular calcification. The patient was accordingly scheduled for

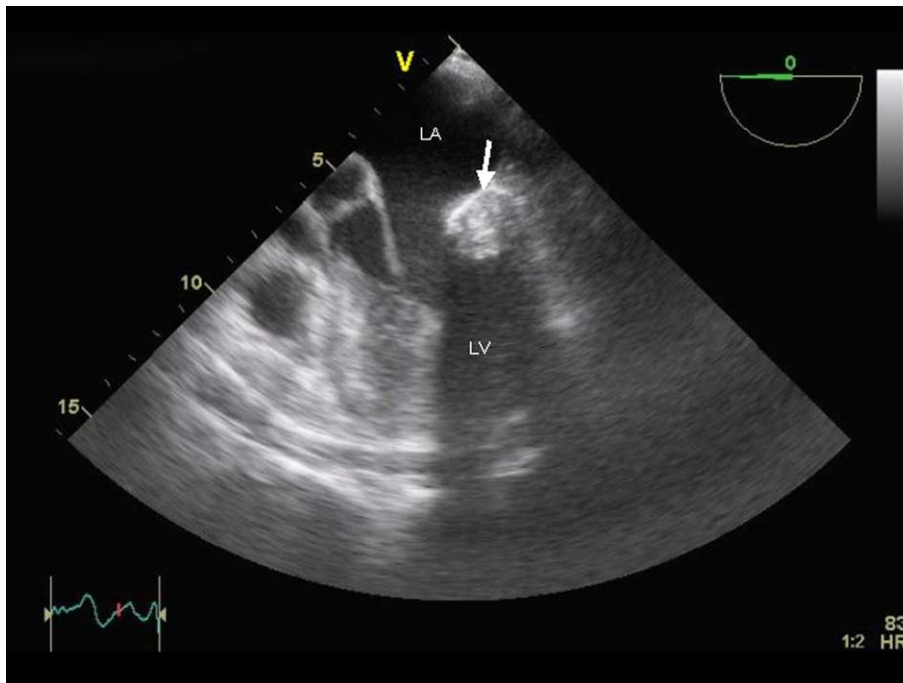


Figure 2 Transesophageal echocardiography showing an inhomogeneous echodense mass (3×2 cm) attached to the posterior mitral annulus. LA, left atrium; LV, left ventricle.

surgery. Because of age and multiple risk factors for atherosclerosis, coronary angiography was performed preoperatively. No significant coronary artery disease was revealed.

During surgery, a large calcified atheroma cyst was observed, originating from the posterior mitral annulus at level P2 with extension to the left atrial and ventricular wall, filled with caseous material (Fig. 3). After detachment of the posterior leaflet, the mass was removed together with the corresponding segment of P2. The posterior leaflet was reconstructed and reattached to the annulus by means of a sliding plasty. A Physio-ring size 32 was then implanted to reform the annulus (Fig. 4). Histopathology confirmed degenerative atheroma and extensive calcium deposit with some fresh thrombotic material (Fig. 5). No granulocytic infiltrates were observed, some giant cells were seen. Cultures were negative.

During echocardiographic follow-up after surgery (transesophageal echocardiography directly postoperative, transthoracic after 1 week, 2 months and 8 months), mitral valve anatomy and function were normal. The patient had an uncomplicated postoperative course and was discharged in stable condition.

Discussion

Intracardiac masses often are an incidental finding by echocardiography. Differential diagnosis of

left-sided masses includes thrombus, vegetation or tumor, most often myxoma. This report presents a case of an incidental finding by echocardiography of a left sided intracardiac mass caused by massive mitral annular calcification.

Mitral annular calcification is a chronic, degenerative process of the mitral valve fibrous ring, primarily involving the posterior annulus. It is an expression of atherosclerosis, with identical risk factors as cardiovascular disease.^{1,2} Mitral annular calcification is observed at autopsy in 3–8% of the population³ and rarely (0.6%) has a large atherosclerotic burden with central “caseous” necrosis resembling a tumor. The incidence of this finding is 0.6% in patients with mitral annular calcification.⁴

Transthoracic echocardiography provides an excellent noninvasive diagnostic technique for detection of intracardiac masses.^{5,6} However, in some patients, limited acoustic viewing restricts diagnostic accuracy. Transthoracic ultrasound images (Fig. 1) of this patient were of insufficient quality to adequately assess the origin and composition of the mass. The transesophageal approach has additional value since it improves visualization of cardiac structures, particularly if located posteriorly. Moreover, transesophageal echocardiography has been demonstrated superior in assessing intracardiac origin of a mass, consistency and mobility.⁵ Echographic differential diagnosis of the mass in this patient included primary or secondary cardiac tumor, vegetation, calcified

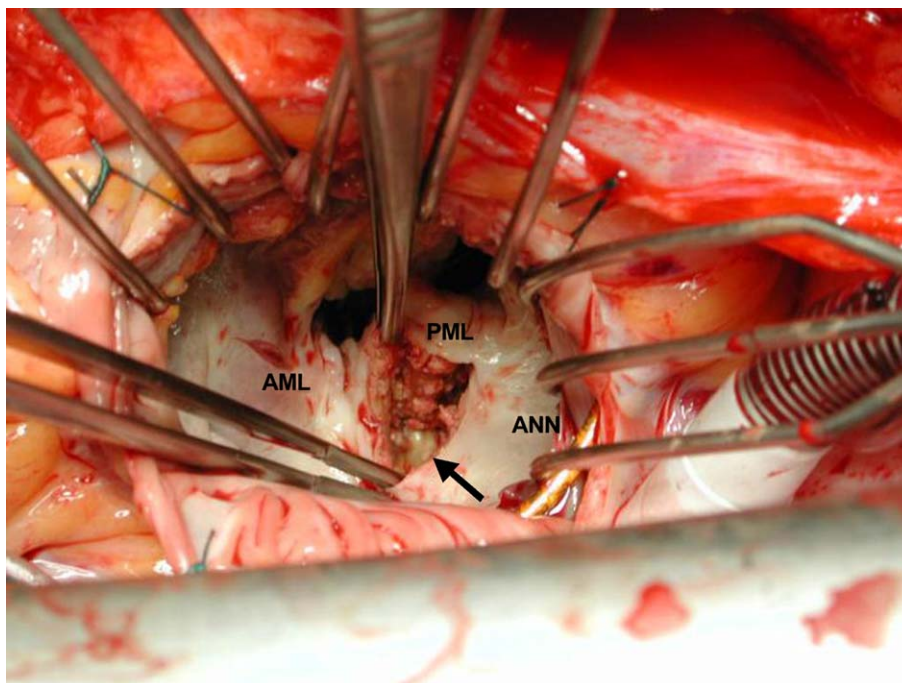


Figure 3 Perioperative illustration of the cystic mass (arrow) seen through the right atrium. Anterior mitral leaflet (AML), posterior mitral leaflet (PML), annulus mitralis (ANN).

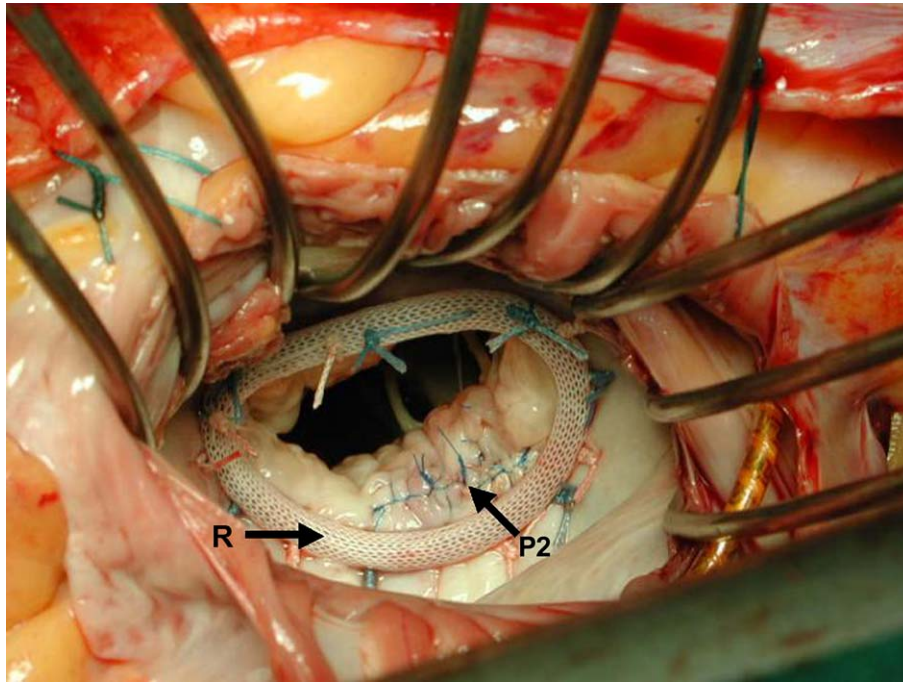


Figure 4 Perioperative illustration of the mitral valve after repair. R, the Physio 32 mm ring; P, the posterior mitral leaflet after partial resection.

thrombus, and extensive mitral annular calcification. The most common intracardiac tumor is a thrombus, but the density of the mass in the current patient suggested calcifications. In the

absence of mitral valve regurgitation, fever, positive blood cultures or infectious laboratory examinations, valvular endocarditis was unlikely. Clinical signs of malignancy were absent. The most

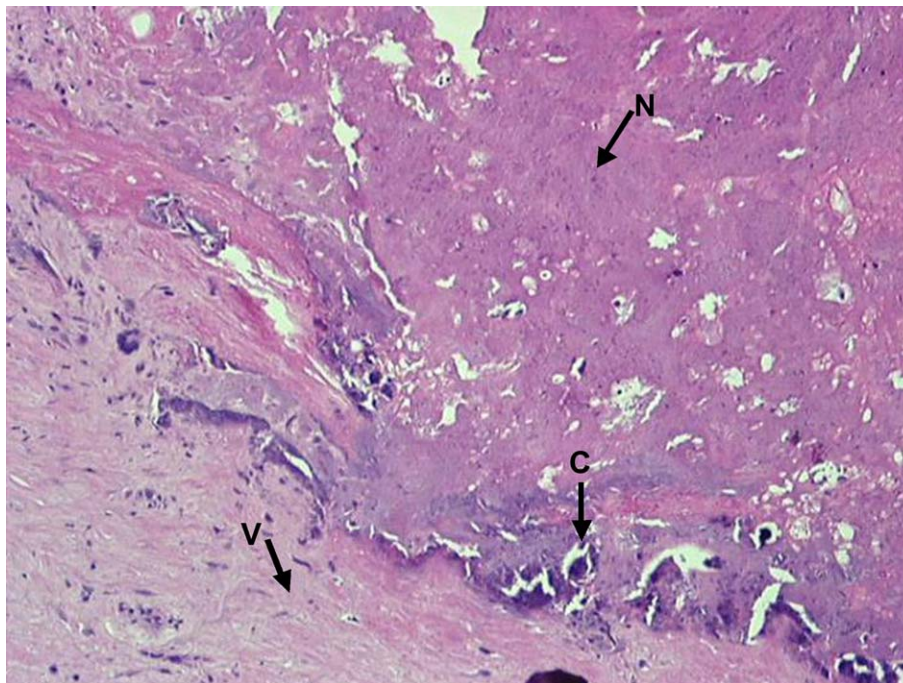


Figure 5 Microscopic examination of caseous material (hematoxylin and eosin, magnification 20 \times). Arrows point out extensive areas of necrosis (N), calcification (C) and some native valve tissue (V).

likely diagnosis was either myxoma or caseous mitral annular calcification. The echodense round, lobular aspect and the mobility of the mass were characteristics favoring the diagnosis of myxoma, although myxomas seldom originate from valves. Despite adequate preoperative imaging, final diagnosis in this patient required surgical exploration and histologic confirmation.

Massive mitral annular calcification is relatively rare and can easily be mistaken for tumor or thrombus.^{7,8} Specific echocardiographic features of caseous mitral annular calcification favor the diagnosis: a large, round echodense, mass in the posterior periannular mitral region, inhomogeneous with central echolucent areas of necrosis. Although mitral annular calcification is often an incidental finding, it has been associated with complications. Rarely, mitral annular calcification is complicated by secondary infection, arrhythmias, mitral regurgitation or stenosis. Several studies have demonstrated an association between mitral annular calcification and stroke.^{9–11} In particular, mitral annular calcification is detected by echocardiography in 25% of patients with stroke.¹² Moreover, Benjamin et al. followed 160 individuals with mitral annular calcification over 8 years in the Framingham cohort.¹² Stroke occurred in 13.8% of patients with mitral annular calcification as compared to 5.1% in the control group. After adjustment of risk factors for cerebrovascular disease, a relative risk of 2.1 to develop stroke was demonstrated.

The mechanism for stroke is unclear. Stein et al. and Malaterre et al. demonstrated thrombus formation on mitral annular calcification with embolization of thrombus.^{10,11} Embolization of small calcified parts of mitral annular calcification is another possible mechanism. Thus, based on the relatively increased risk of embolization, surgery may be the preferred treatment option, especially if the valve can be repaired. Harpaz et al. however, followed 13 patients with mitral annular calcification and caseous necrosis over 3.8 years; all patients were treated conservatively and stroke did not occur in any of the patients.⁴

Based on these scarce data in the literature, there appears currently no consensus on the optimal treatment of caseous mitral annular

calcification, and further studies are needed to resolve this issue.

Conclusion

In conclusion, a calcified tumor connected to the posterior mitral annulus should raise suspicion of a pseudotumor caused by mitral annular calcification. Although the available data in the literature indicate that mitral annular calcification is associated with an increased risk of stroke, the optimal treatment (conservative or surgical) remains to be established.

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