Rare disease

Two cases of massive mitral annular calcification mimicking left atrial neoplasms

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Summary

The authors describe two cases of massive mitral annular calcification, initially picked up on echocardiography and suspected of being neoplastic. Subsequent evaluation by CT scanning confirmed the location, aetiology, structure and diagnosis. Both cases demonstrated large masses, with calcification of varying density. This is likely explained by the presence of the previously reported amorphous caseous material demonstrated to be present within such mass structures. Such a feature is described as caseous degeneration. Both patients described have been managed conservatively with medical therapy, predominantly due to their age and general frailty.

BACKGROUND

These cases were rather extreme examples of a more common echocardiographic feature. The important lesson is that such features should not be confused with intracardiac tumours and how they can be better imaged using CT imaging.

Mitral annular calcification (MAC) is relatively common, with a prevalence of 5–25% in patients over 40 years of age,^{1 2} which increases to more than 40% in the older.³ It is associated with increased risk of myocardial infarction and cardiac death.² The prevalence of caseous degeneration is reported as being 0.6% in patients with MAC and was between 0.06% and 0.07% in a large series of patients of all ages.⁴ This contrasts with only 2 cases, albeit of an extreme nature, that we have identified from over 44 000 echocardiograms (0.00005%) in the last 11 years.

CASE PRESENTATION

Case 1 A 78-year-old Asian woman presented with extreme shortness of breath and an exercise capacity of a few yards. One year before, she had developed atrial fibrillation with an associated deterioration in her activities. She had been on long-term steroids for rheumatoid arthritis, thyroid supplementation and treatment for raised blood pressure. Three months previously she had experienced an episode of amaurosis fugax which was treated with aspirin. Clinically, she was in rate controlled atrial fibrillation and normotensive at 140/70 mm Hg. The jugular venous pressure was elevated by 5 cm with mild bilateral peripheral oedema. On auscultation, there was an ejection systolic murmur in the aortic area with a pansystolic murmur audible throughout the precordium.

Case 2 An 81-year-old Asian woman presented with shortness of breath and leg swelling of 6 months duration. Clinically she was normotensive and in rate controlled atrial fibrillation. The jugular venous pressure was elevated

by 10 cm and oedema was present to the top of both thighs. Auscultation revealed a harsh systolic and diastolic murmur audible over the precordium. Medical history included treatment for hypertension and a temporo-parietal ischaemic stroke 9 years earlier.

INVESTIGATIONS

Case 1 A chest radiograph showed cardiomegaly and upper lobe blood diversion. An echocardiogram confirmed aortic stenosis with a peak gradient of 76 mm Hg (mean 41 mm Hg) and a calculated, valve area of 0.8 cm². The left ventricle was normal sized and hyperdynamic left ventricle (ejection fraction >80%). The left atrium was significantly dilated (56 mm) with moderate mitral regurgitation. A large mass (2.6 x 2.6 cm on a strict parasternal long-axis view) was noted, that appeared to arise from the posterior mitral valve leaflet (figure 1a). An ECG gated CT scan of the heart was performed as part of transcatheter aortic valve implantation assessment which revealed a densely calcified mass on precontrast images (figure 1b) but with varying degree of density (600–1700 HU) on contrast images (figure 1c). The mass predominantly occupied the posterior part of the valve annulus and pushed the posterior mitral leaflet anteriorly. Biochemistry revealed a normal corrected serum calcium of 2.37 mmol/l with a mildly depressed glomerular filtration rate of 48 ml/min. The serum creatinine (99 mmol/l) and urea (6.0 mmol/l) were normal.

Case 2 The chest radiograph showed a 2.5 cm ovoid opacity projecting over the left lower zone and seen through the cardiac silhouette (figure 2a). An ECG demonstrated good global left ventricular function. The left atrium was enlarged at 46 mm. In addition to heavy MAC, a well-circumscribed region of calcification, measuring 4.2 x 2.8 cm, was noted (figure 2b) in the region of the posterior mitral valve attachment with moderate mitral

Figure 1 Echocardiogram in left parasternal view (a) shows a mass in the posterior mitral region. A precontrast axial CT short image (b) shows a large highly calcified mass with the long-axis (c) contrast image showing its precise location (arrows) and variable density (high as large arrow and low as black star).

regurgitation. Again, an ECG gated CT scan confirmed a variable but high density mass along the posterior mitral annulus, confined anteriorly by the posterior mitral leaflet (figure 2c). The renal function was depressed with a glomerular filtration rate of 25 ml/min and a raised serum creatinine (172 mmol/l) and urea (17.6 mmol/l). The serum calcium was not performed in this case.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for both these masses, that each measured about 1.5 cm in a long-axis and 4–5 cm in a short-axis planes, was primarily that of an intracardiac neoplasm. This might include a primary cardiac tumour, the commonest being a myxoma. Other types of tumour, less commonly seen include fibroelastomas, rhabdomyomas, teratomas (usually in infants), angiosarcomas, fibrosarcomas and rhabdomyosarcomas. Secondary metastases can also frequently involve the heart. Other considerations include organised thrombus, abscesses or vegetations in association with endocarditis.

TREATMENT

Case 1 declined further investigations on the grounds that she would not contemplate cardiac surgery. She continues

on medical treatment. Case 2 was also managed conservatively, predominantly due to her general frailty and age.

OUTCOME AND FOLLOW-UP

Both patients are still alive 1 year after they were first investigated and continue to be seen as outpatients with little further deterioration in their initial symptomology.

DISCUSSION

MAC is relatively common, with a prevalence of 5–25% in patients over 40 years of age,² which increases to more than 40% in the older.³ It is associated with increased risk of myocardial infarction and cardiac death.² The prevalence of caseous degeneration is reported as being 0.6% in patients with MAC and was between 0.06% and 0.07% in a large series of patients of all ages.⁴ This contrasts with only 2 cases, albeit of an extreme nature, that we have identified from over 44 000 ECG (0.00005%) in the last 11 years. Both our patients were older Asian women and both had been treated for hypertension. One of the largest studies published, reported on 14 patients with caseous mitral valve calcification; all 14 were older and all hypertensive.⁵ In most case reports and the few small published series, the subjects are for some unexplained reason predominantly

Figure 2 Chest x-ray (a) shows dense calcification in the mitral area (arrows) which on a magnified echocardiographic four chamber view (b) is demonstrated to be an ellipsoid mass (arrows). A contrast short-axis CT scan (c) shows the location of the large calcified mass (arrows) in the region of posterior mitral annulus, outlined on the inside by the posterior mitral leaflet (small arrows) and containing high and low density regions.

older women.⁶ At presentation, both our patients were in atrial fibrillation, were normocalcaemic and had normal inflammatory markers. In common with a number of publications,^{7–9} we had an initial suspicion that these intracardiac masses might represent intracardiac tumours. We observed interference with both the mitral valve and the left ventricular outflow tract by direct physical obstruction to blood flow. In some reports, the calcification has been described as extending into the myocardium¹⁰ with resultant dysfunction of the subvalvular apparatus and a restrictive cardiomyopathy.¹¹ Both mitral regurgitation and mitral stenosis are described as consequences of this process.¹²

In recent years, mitral caseous calcification has been more clearly characterised using CT and MRI.^{8 13–17} Non-contrast CT typically shows a large calcified mass in the region of posterior mitral annulus extending to the adjacent mitral valve and myocardium.⁸ On contrast enhanced CT, the central part appears less hypodense due to the caseous toothpaste-like material contained within the denser calcified peripheral rim.^{6 13 15} MRI has shown low signal on both T1 and T2 weighted images due to calcification but is inferior in showing the calcification itself directly.¹³ However, these different imaging modalities are clearly

complimentary,¹⁸ with echocardiography initially detecting these mass abnormalities and defining their functional significance. MRI has a role in excluding other potential aetiologies and will confirm the presence of calcification but, compared to CT imaging, less optimally discriminates transformation into caseous necrosis.¹⁶

When associated with mitral valve dysfunction, cardiac surgery to excise the mass and replace the valve, with or without ring annuloplasty, appears to be a reasonable option.^{12 19} A surprising feature of this condition includes reports of spontaneous resolution;^{5 20} possibly due to resorption in a ‘necrotic liquefaction’ process or via ‘extrusion’ of the contained semiliquid material.

A limitation to both cases is that the serum phosphate and parathyroid hormone levels were not measured.

In conclusion, although the mitral annulus is a common site for calcification in the older, more rarely large calcified intracardiac masses can be mistaken for neoplasms on echocardiography. In itself a benign condition, the physical presence of a large mass has significant implications to the functioning of the mitral valve and in some cases to left ventricular outflow. CT scanning provided an invaluable tool for diagnosis of such intracardiac masses, and to assess the influence on adjacent structures. In many cases

of caseous calcification, multi-modality imaging, including cardiac MRI, is often employed.

Learning points

- MAC is an unusual pathology.
- Appearances may be mistaken for intracardiac neoplasms.
- CT scanning provides helpful images to differentiate the diagnosis.
- Massive MAC may cause obstructive issues.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Harpaz D**, Auerbach I, Vered Z, *et al.* Caseous calcification of the mitral annulus: a neglected, unrecognized diagnosis. *J Am Soc Echocardiogr* 2001;**14**:825–31.
2. **Fox CS**, Vasan RS, Parise H, *et al.*; Framingham Heart Study. Mitral annular calcification predicts cardiovascular morbidity and mortality: the Framingham Heart Study. *Circulation* 2003;**107**:1492–6.
3. **Barasch E**, Gottdiener JS, Larsen EK, *et al.* Clinical significance of calcification of the fibrous skeleton of the heart and atherosclerosis in community dwelling elderly. The cardiovascular health study (CHS). *Am Heart J* 2006;**151**:39–47.
4. **Novaro GM**, Griffin BP, Hammer DF. Caseous calcification of the mitral annulus: an underappreciated variant. *Heart* 2004;**90**:388.
5. **Deluca G**, Correale M, Ieva R, *et al.* The incidence and clinical course of caseous calcification of the mitral annulus: a prospective echocardiographic study. *J Am Soc Echocardiogr* 2008;**21**:828–33.
6. **Fernandes RM**, Branco LM, Galrinho A, *et al.* Caseous calcification of the mitral annulus. A review of six cases. *Rev Port Cardiol* 2007;**26**:1059–70.
7. **Teja K**, Gibson RS, Nolan SP. Atrial extension of mitral annular calcification mimicking intracardiac tumor. *Clin Cardiol* 1987;**10**:546–8.
8. **De Conti F**, Corbetti F, De Conti G, *et al.* Unusual tumor-like calcification of the mitral annulus: diagnosis and tissue characterization by ultrasound, computed tomography and magnetic resonance imaging. *Ital Heart J* 2005;**6**:160–3.
9. **de Vrey EA**, Scholte AJ, Krauss XH, *et al.* Intracardiac pseudotumor caused by mitral annular calcification. *Eur J Echocardiogr* 2006;**7**:62–6.
10. **Butz T**, van Bracht M, Meissner A, *et al.* Mitral valve disease as well as uncommon extensive epi-pericardial and intramyocardial calcification secondary to massive mitral annular calcification. *Eur J Echocardiogr* 2010;**11**:85–8.
11. **Salisbury AC**, Shapiro BP, Martinez MW. Extensive myocardial and mitral annular calcification leading to mitral regurgitation and restrictive cardiomyopathy: An unusual case of caseous calcification of the mitral annulus. *J Cardiovasc Comput Tomogr* 2009;**3**:351–3.
12. **Alkadhi H**, Leschka S, Prêtre R, *et al.* Caseous calcification of the mitral annulus. *J Thorac Cardiovasc Surg* 2005;**129**:1438–40.
13. **Vanovermeire OM**, Duerinckx AJ, Duncan DA, *et al.* Caseous calcification of the mitral annulus imaged with 64-slice multidetector CT and magnetic resonance imaging. *Int J Cardiovasc Imaging* 2006;**22**:553–9.
14. **Monti L**, Renifilo E, Profili M, *et al.* Cardiovascular magnetic resonance features of caseous calcification of the mitral annulus. *J Cardiovasc Magn Reson* 2008;**10**:25.
15. **Zeina AR**, Makhoul N, Nachtigal A. Mitral annulus caseous calcification imaged with 64-slice MDCT. *Acute Card Care* 2009;**11**:121–2.
16. **Blankstein R**, Durst R, Picard MH, *et al.* Progression of mitral annulus calcification to caseous necrosis of the mitral valve: complementary role of multi-modality imaging. *Eur Heart J* 2009;**30**:304.
17. **Providência R**, Botelho A, Mota P, *et al.* Cardiac mass in a patient with colorectal cancer: “not all that glitters is gold!”. *World J Cardiol* 2011;**3**:98–100.
18. **Gulati A**, Chan C, Duncan A, *et al.* Multimodality cardiac imaging in the evaluation of mitral annular caseous calcification. *Circulation* 2011;**123**:e1–2.
19. **Minardi G**, Manzara C, Pulignano G, *et al.* Caseous calcification of the mitral annulus with mitral regurgitation and impairment of functional capacity: a case report. *J Med Case Reports* 2008;**2**:205.
20. **Gramenzi S**, Mazzola AA, Tagliaferri B, *et al.* Caseous calcification of the mitral annulus: unusual case of spontaneous resolution. *Echocardiography* 2005;**22**:510–3.

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