CASE REPORT

Severe mixed mitral valve disease due to massive mitral annular calcification: A case report and literature review

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Abstract

We present an unusual case of massive mitral annular calcification (MAC) leading to severe mixed mitral valve disease, viz severe mitral regurgitation and severe mitral stenosis. To our knowledge, severe mixed mitral valve disease secondary to MAC is extremely rare. The patient (a 65-year-old lady) presented with worsening shortness of breath and signs of congestive heart failure. Echocardiographic examination revealed massive mitral annular calcification. Despite the massive annular calcification, she had experienced neither embolism nor endocarditis in the past. Because of severe symptomatic mitral regurgitation and mitral stenosis, surgery was advised; however, the patient declined it.

Keywords: Mitral annular calcification, degenerative mitral valve disease, mitral regurgitation, mitral stenosis.

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Introduction

Mitral annular calcification (MAC) is a chronic, degenerative process of the mitral valve fibrous ring seen in approximately 10% of patients over 50 years of age. Some of this degenerative change has prototypical echocardiographic findings termed 'liquefaction necrosis' or 'caseous calcification', where a mass with a central echolucent area is seen.[1] Massive mitral annular calcification may simulate an intra-cardiac tumour.[2] It can lead to mitral regurgitation, mitral stenosis or a combination of both. Furthermore, it has been associated with an increased risk of endocarditis, atrial fibrillation and stroke.[1,3,4] Management depends on the presenting clinical scenario including the degree of symptomatic valvular abnormality and the patient's comorbidities.[5] Here, we report an unusual case of massive mitral annular calcification leading to severe mitral regurgitation and severe mitral stenosis due to obstruction at the annular level.

Case Report

This case report was given an exemption by the hospital's ethical review committee (ERC No. 2020-4737-10744).

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A 65-year-old lady with a history of hypertension presented with worsening shortness of breath at the Aga Khan University Hospital, Karachi, in October, 2019. She had a holosystolic murmur consistent with mitral regurgitation with clinical and radiological signs of congestive heart failure. A 12-lead electrocardiogram (ECG) showed nonspecific changes and troponins were negative. Renal function was mildly impaired with a creatinine of 1.49 mg/dl (Normal range 0.59 – 1.04 mg/dl), while serum calcium and parathyroid hormone levels were normal. Transthoracic 2D echocardiography revealed massive mitral annular calcification of both the anterior and posterior mitral leaflets (Figure 1a) with elevated gradients

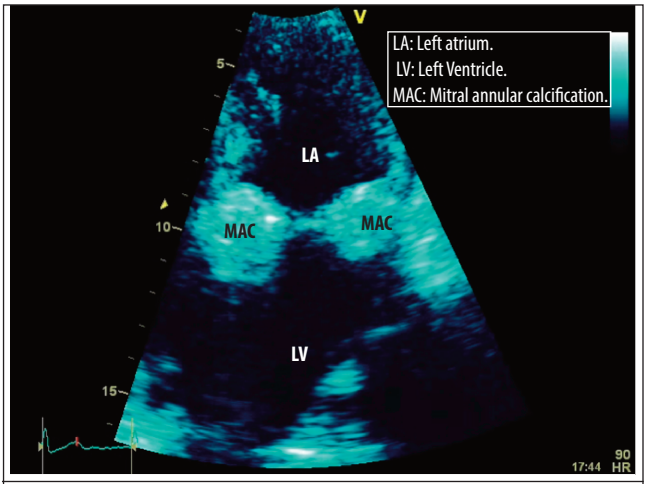

Figure-1 a: Massive mitral annular calcification on 2-D echo.

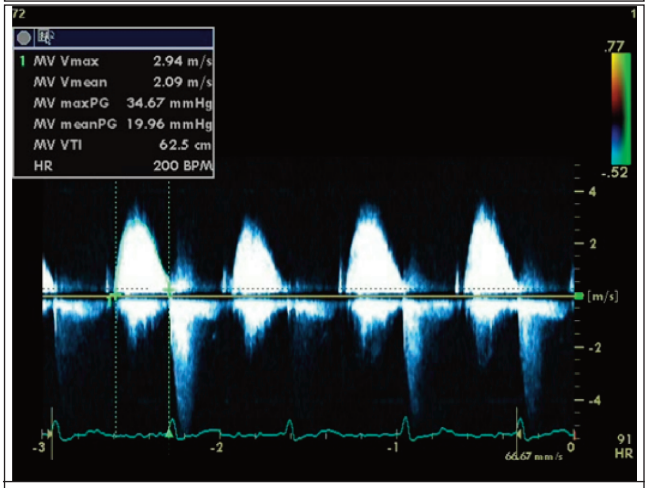

Figure-1 b: Elevated gradients across mitral valve on Doppler.

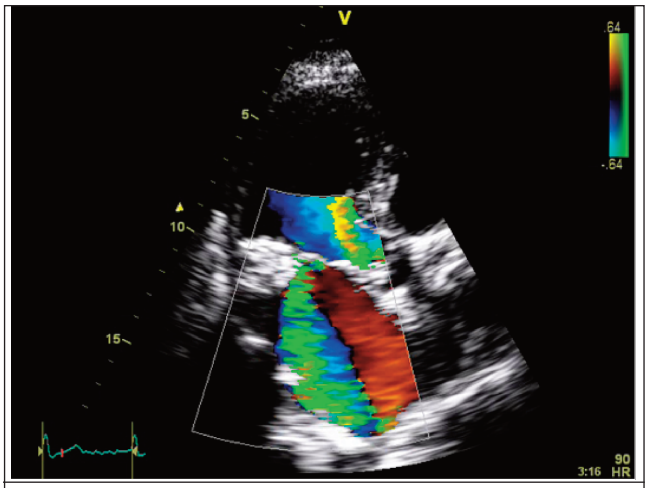

Figure-1 c: Mitral regurgitation jet on colour flow Doppler.

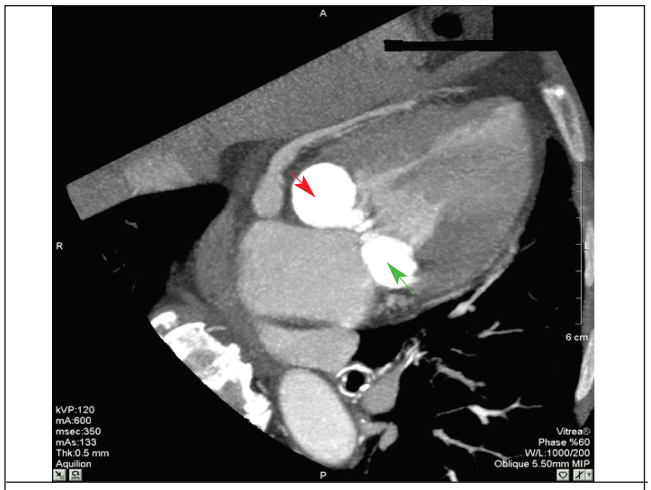

Figure-2 a: Cardiac Computed tomography, oblique sagittal view through the left ventricle and left atrium shows dense mitral annulus calcification extending centrally (Red arrows). The calcification does not involve the chordae tendinae or papillary muscles (Green arrow).

across the mitral valve (mean and peak diastolic gradients were 19 and 38mmHg, respectively) and severe mitral regurgitation (Figure 1b & 1c). Left ventricular ejection fraction was normal.

The patient's condition improved with diuresis and beta blockade. Cardiac computerised tomography (CT) was performed to evaluate the extent of calcification and to better assess if the adjoining myocardium or the papillary apparatus was involved. No extension was noted beyond the mitral annulus. (Figure 2a & 2b) Surgical excision with valve replacement was offered, but the patient declined.

Discussion

Calcification of the mitral valve annulus is a chronic degenerative process. There are multiple proposed pathophysiological mechanisms; MAC is seen in elderly

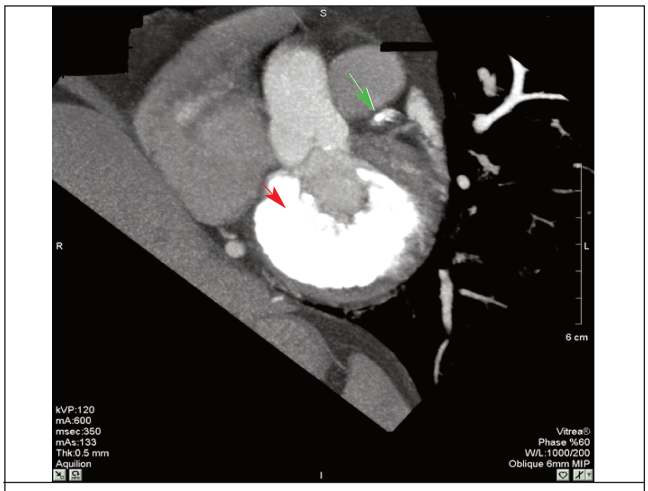

Figure-2 b: Cardiac computed tomography, short axis view at the level of the mitral annulus shows dense curvilinear calcification (Red arrow). Incidentally there is evidence of calcific coronary artery disease (Green arrow).

patients and in people with other cardiovascular risk factors. It is, therefore, believed to have a shared pathogenesis with atherosclerosis. It is also seen in people with chronic kidney disease and disordered calcium metabolism. Several epidemiologic studies describe MAC.[6] MAC is more commonly associated with mitral regurgitation and less with mitral stenosis.[4] Various studies have shown differing incidence of mitral stenosis in this setting. In a study conducted by Bouvier et al, the occurrence of moderate-to-severe degenerative MS was as high as in 34% of patients with mitral calcific deposits, including 12% with severe to very severe MS as detected by multi-detector CT in patients referred for transcatheter aortic valve implantation (TAVI) due to aortic stenosis.[7] MAC is also associated with increased resting gradient across mitral valve and as reported by Betrand et al it is prognostically significant that 10-year survival rate is only 11% in patients with MAC having mean transmitral gradient of >10mmHg.[8] A possible explanation for the aetiology of mitral regurgitation in the setting of annular calcification is reduced mobility of the mitral valve and chordae due to the calcification. There may also be increased traction on the chordae, leading to chordal dysfunction or rupture with secondary mitral regurgitation. Additional explanations include loss of the 'sphincter' action of the mitral valve in systole and a failure to contract at end diastole.

There are a number of mechanisms postulated for mitral stenosis in MAC. A severely calcified mitral annulus restricts leaflet mobility. This effect appears to be greater in patients with significant involvement of the anterior mitral leaflet (AML), where the calcification displaces the 'hinge point' of the mitral leaflet. Calcification can also limit the function of the chordae. Our patient was noted to have severe

calcification of both AML and PML.

The occurrence of mixed disease is seen in a series where patients underwent surgery for mitral valve dysfunction associated with MAC. For example, Uchimuro et al, in 2016, evaluated 61 patients with MAC who underwent surgery for mitral valve disease. Of these, 19 patients 19(31%) were reported to have mixed mitral stenosis and regurgitation. This high incidence was not seen in echocardiography-based studies, suggesting that mixed disease is a rare entity seen in patients with particularly severe MAC causing valvular dysfunction.[9]

No treatment is required for MAC that does not obstruct mitral inflow or cause regurgitation, unless severe. Conservative management includes aggressive cardiovascular risk factor modification and consideration of anticoagulation given the risk of thrombus formation on the mitral annular region with potential for embolization. In patients with renal dysfunction, disordered calcium metabolism should be managed appropriately, if necessary.

If MAC is associated with significant mitral regurgitation or stenosis, the treatment is surgical excision with valve replacement or repair, if possible. Surgery for mitral valve disease in the setting of MAC is a challenge with significant risk for stroke and cardiac rupture.[10] Newer techniques utilising transcatheter mitral valve replacement are currently being explored in patients deemed unsuitable for surgery but it may have its own adverse effects.[10]

Conclusion

This is an unusual case of massive mitral annular calcification leading to severe mitral regurgitation and severe mitral stenosis due to obstruction at the annular level.

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Conflict of interest: None.

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