

Recurrent ST-elevation myocardial infarction: a case report of a rare complication of caseous mitral annular calcification

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Background

Caseous mitral annular calcification (MAC) is an under-diagnosed division of calcific mitral valve disease that has recently been reported to have increased propensity for embolic disease. Early recognition of this entity as a cause of embolic disease can lead to prevention of occlusive vascular disease and long-standing complications.

Case summary

We present the case of a patient with end-stage renal disease who presented for evaluation of chest pain and was found to have ST-segment myocardial infarction. Despite thrombectomy and stenting, he had multiple recurrent events, and imaging evaluation demonstrated caseous MAC with mobile components. He was taken for surgical replacement of the mitral valve, with pathology confirming diagnosis.

Discussion

Caseous MAC may represent an increased risk of embolic disease. Better understanding of this pathology and its propensity for embolic disease will be important to best determine treatment plans and timing of operative intervention.

Keywords

Mitral annular calcification • Embolic disease • Valvular surgery • Case report

ESC Curriculum

2.2 Echocardiography • 3.2 Acute coronary syndrome • 4.4 Mitral stenosis

Learning points

- Caseous mitral annular calcification is an under-recognized disease that can develop rapidly and pose significant risk of embolic disease, both from calcific components and thrombus formation.
- Intervention may be warranted for those with evidence of embolic disease, but the role of prophylactic intervention is unclear at this time.

Introduction

Caseous mitral annular calcification (MAC) is a variant of degenerative calcific changes of the mitral valve that may present with sterile abscesses of the valvular apparatus.¹ Previous studies have suggested the prevalence to be less than 1% of all MAC; however,

autopsy studies have suggested higher prevalence, which may be due to difficulty with diagnosis, variable presentation on imaging, and lack of physician awareness.² Caseous MAC has traditionally not been considered to have high embolic risk.^{3,4} However, case reports have suggested that in younger patients, particularly those

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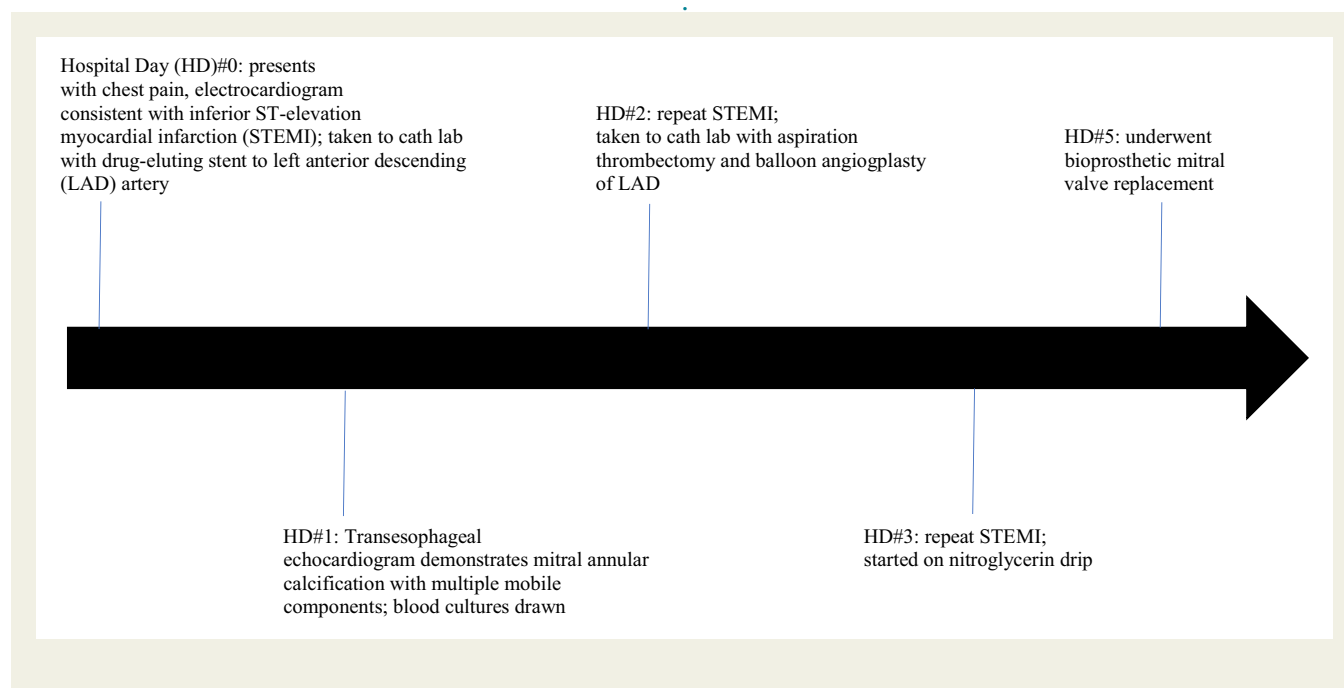
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with renal disease, this may be a potentially under-recognized clinical diagnosis that can present with embolic phenomenon.^{5,6} We present the case of a young male presenting with recurrent ST-elevation myocardial infarction due to embolic calcific disease.

Timeline



plaque rupture or embolic event most likely on this patient's differential diagnosis.

Given the concern for coronary occlusion, patient was taken to the cardiac catheterization lab. Initial angiogram ([Video 1](#)) demonstrated thrombotic occlusion of the distal left anterior descending artery, which was a large wrap-around vessel supplying the anterolateral and portions of the inferior wall of the left ventricle. Rheolytic thrombectomy was attempted with successful retrieval of some

Case presentation

A 37-year-old male presented to the emergency department (ED) with chest pain beginning during his dialysis session earlier in the day. The pain was left sided, without radiation, and without changes in quality or severity with positional manoeuvres. He completed his dialysis session, but the pain persisted, prompting presentation to ED.

Past medical history was significant for end-stage renal disease, uncontrolled hypertension, and type 1 diabetes mellitus. He had no history of anginal symptoms and was active without limitations. There was no known family history of early coronary artery disease, sudden cardiac death, or cardiomyopathies.

At presentation, he was hypertensive (blood pressure 192/126 mmHg) and tachypnoeic, saturating 98% on 3L via nasal cannula. On exam, he appeared uncomfortable and was noted to be tachycardic with grade 2/6 systolic murmur at the apex without significant radiation. There was jugular venous distention to the angle of the mandible and crackles noted in the bilateral lung bases. Electrocardiogram was performed, demonstrating infero-lateral ST-segment elevations ([Figure 1](#)).

The differential diagnosis for ST-segment elevations includes: acute coronary event due to plaque rupture, stress-induced cardiomyopathy, coronary vasospasm, embolic coronary event, pericarditis, myocarditis, and pulmonary embolism. Despite his young age, the acuity of symptoms, distribution of ST-segment abnormalities, and presence of multiple cardiac risk factors made acute coronary occlusion due to

thrombus, and patient was started on intravenous GIIb/IIIa inhibition in addition to oral P2Y12 inhibitor following 3.25 × 26 mm Resolute Onyx (Medtronic, Warsaw, IN, USA) stent placement.

Transthoracic echocardiogram was performed per European Society of Cardiology guidelines, which recommend early echocardiography for all patients presenting with ST-elevation myocardial infarction to assess left ventricular (LV) systolic function, identify potential complications of obstructive coronary disease, and identify potential sources.⁷ This demonstrated preserved LV performance with apical hypokinesia. There was no evidence of LV thrombus, but the mitral valve demonstrated echo-bright, mobile densities on the ventricular side of the anterior mitral leaflet as well as a heterogeneous echodensity on the atrial side of the posterior mitral annulus ([Figure 2](#)). The brightness suggested calcification, but mobility of the lesions was also concerning for vegetation. Blood cultures were drawn, which remained negative. No leucocytosis was noted and the patient remained afebrile without signs or symptoms or infection. He denied any previous infectious symptoms, intravenous drug use, or indwelling lines within the last year.

Transoesophageal echocardiogram was performed, which confirmed the presence of at least three discrete bright, echo-dense lesions involving both leaflets of the mitral valve, the largest of which measured 0.9 cm by 0.8 cm ([Video 2, Supplementary material online, Video S1, Figure 3](#)). One lesion was noted to protrude into the LV outflow tract, and all lesions were highly mobile at their attachment site. Valvular function was not affected, with only mild mitral stenosis and regurgitation, and no other valvular disease.

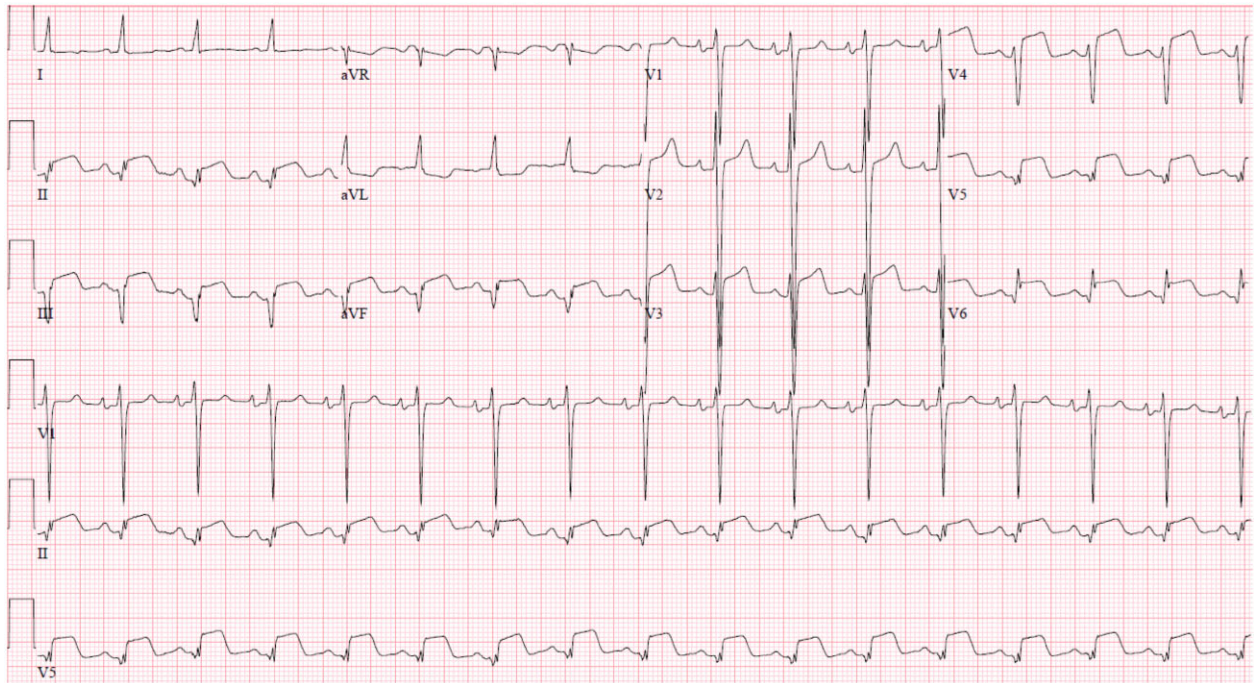
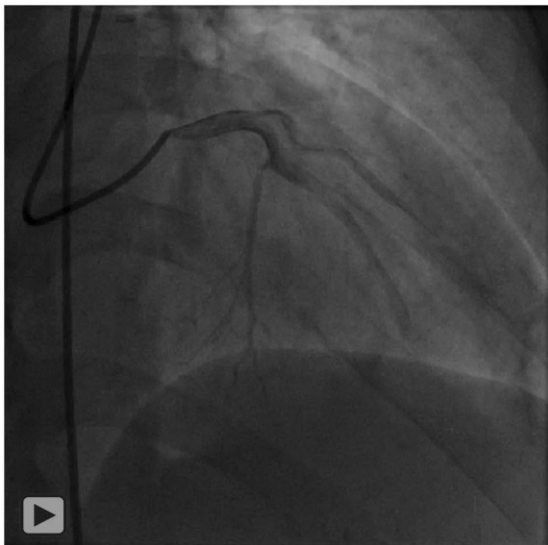


Figure 1 Initial electrocardiogram demonstrating ST-segment elevations in infero-lateral leads.



Video 1 Angiogram demonstrating distal left anterior descending artery occlusion.

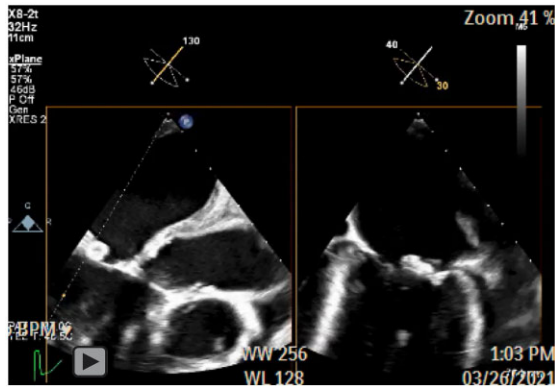
Following initial percutaneous coronary intervention, the patient had improvement in his ST segments and resolution of chest pain. However, 48 h after initial presentation, he had recurrent chest pain with associated diaphoresis and tachycardia. Electrocardiogram again demonstrated ST-segment elevation in the inferolateral leads, and he was taken back to the cardiac catheterization lab. The previously



Figure 2 Transthoracic echocardiogram demonstrating calcification of mitral valve.

placed stent was patent, but distal to this there was an occlusive lesion. Aspiration thrombectomy was performed with removal of thick, white material that appeared consistent with calcification. Serial balloon dilations were made and overlapping stent was placed to achieve thrombolysis in myocardial infarction (TIMI) III flow in the vessel.

The patient had two additional events during the next 48 h, which were managed conservatively with nitroglycerine drip and antiplatelet agents. After multidisciplinary discussion, decision was made to proceed with mitral valve replacement with patch reinforcement of posterior annulus. Prior to this, despite his lack of neurologic symptoms on regular exams, the patient underwent computed



Video 2 Transoesophageal echocardiogram (biplane) demonstrating mobile calcification of mitral valve.

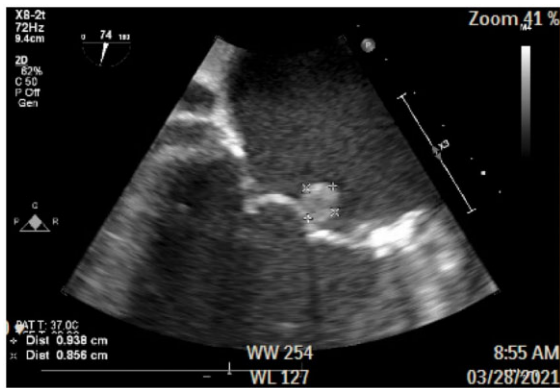
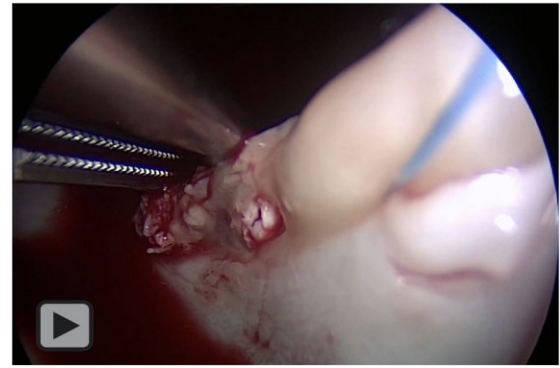


Figure 3 Transoesophageal echocardiogram demonstrating size of mobile calcific lesion on mitral valve.

tomography scan to rule out large territory stroke or intracranial haemorrhage per American Association of Thoracic Surgeons (AATS) guidelines.⁸ Previous studies have not demonstrated any significant difference in mortality between mechanical and bioprosthetic valves in patients with end-stage renal disease undergoing mitral valve replacement, and those with bioprosthetic mitral valves tend to have lower rates of perioperative complications due to lower bleeding risk.^{9,10} Given this, his multiple co-morbid conditions, and concerns with anticoagulation compliance, decision was made to proceed with a 27 mm Epic bioprosthetic valve in this patient.

Intraoperatively, gelatinous, 'toothpaste-like' material was noted (Video 3) and surgical pathology confirmed fibrocalcific disease of the valve. Histologic examination demonstrated fibrosis and exophytic calcification without any evidence of inflammation or inter- or intracellular organisms (Figure 4), consistent with negative blood cultures and lack of clinical symptoms suggestive of infective endocarditis.

The patient was discharged home after an unremarkable post-operative course. At 3-month follow-up, he reported no recurrent episodes of chest pain or functional limitations and was able to



Video 3 Intraoperative video demonstrating mitral annular calcification.

exercise at baseline level. Echocardiogram demonstrated preserved LV systolic function despite apical akinesis, with normally functioning bioprosthetic valve gradients and no evidence of significant calcification on or around valve leaflets.

Discussion

Caseous MAC generally affects older patients with longstanding hypertension, but younger patients with renal disease or disorders of calcium metabolism may also be affected.⁴ In these younger patients, the development of mitral calcification and associated mobile components can occur rapidly and should be high on the differential in the correct clinical context regardless of prior imaging.¹¹

Previous studies have suggested caseous MAC is benign and does not carry the same association with embolic disease as traditional MAC.^{3,4} However, as identification of caseous MAC has improved, there have been increasing reports of embolic events associated with caseous MAC, with prevalence as high as 20%, which is higher than reported prevalence of embolic disease with traditional MAC.⁵ Although MAC itself serves as a nidus for thrombus formation, embolization of loosely adherent caseous necrotic debris or fistulization of caseous materials into the left atrium or left ventricle may account for the proposed increased risk of embolic disease with caseous MAC.^{5,6}

Despite these findings, there is no current recommendation to empirically treat caseous MAC to prevent embolic phenomenon. Current practice based on previous case reports appears to rely heavily on operative management following established embolic disease.^{12,13} Operative intervention itself can pose risk of further embolization, and as caseous MAC becomes more recognized, new operative techniques have been described to help mitigate this risk.¹⁴

Due to these differences in aetiology of embolic risk, the differentiation between MAC and caseous MAC has important therapeutic considerations. Our patient is unique in that while subsequent events were the result of embolic calcific disease, his original presentation was due to suspected thrombus. There was no evidence of atrial defect, peripheral thrombus, ventricular thrombus, or atrial arrhythmias

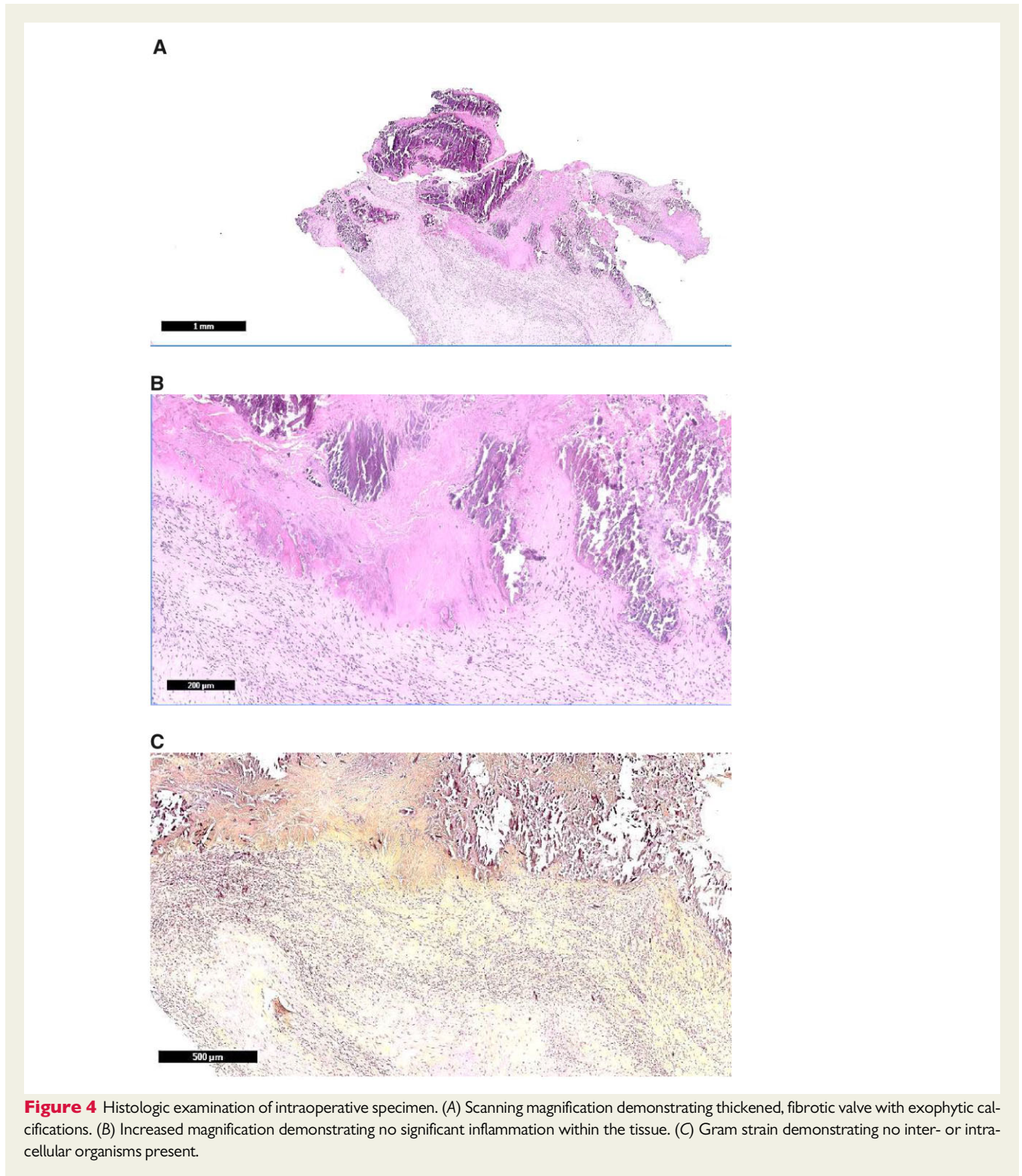


Figure 4 Histologic examination of intraoperative specimen. (A) Scanning magnification demonstrating thickened, fibrotic valve with exophytic calcifications. (B) Increased magnification demonstrating no significant inflammation within the tissue. (C) Gram stain demonstrating no inter- or intra-cellular organisms present.

to suggest alternative mechanism for thrombus formation, and we propose that this suspected thrombus originated from his calcific mitral valve. To the best of our knowledge, this is the first report of caseous MAC associated with suspected thrombus formation and distal embolization. Our case is also unique in that no other common

sources of cardiac emboli were noted, and the patient had recurrent episodes of coronary occlusions in the same vessel due to the angle of left coronary ostium.

Caseous MAC itself is not currently an indication for surgery, despite evidence for embolic risk. In those patients with evidence of

embolic disease or large, mobile lesions on the valve with high embolic risk, we believe it would be reasonable to proceed with surgery, extrapolating from current endocarditis guidelines.⁸ In patients with neurologic symptoms, imaging may be warranted to rule out embolic disease to the brain which may be at risk for haemorrhagic conversion during cardiopulmonary bypass. Given the rapid progression and suspected high embolic risk, it may be reasonable to consider empiric elective surgical resection of caseous MAC in patients who are good surgical candidates.⁵

Conclusions

Caseous MAC is an under-recognized disease that can develop rapidly and pose significant risk of embolic disease, both from calcific components and subsequent thrombus formation. We propose that surgical indications should follow those of endocarditis, and there may be a potential role for prophylactic valve intervention in those patients who have developed caseous MAC without symptoms and are good surgical candidates.

Lead author biography



Dr Nikhil Singh is a third-year cardiology fellow at the University of Chicago Medical Center in Chicago, IL, USA. He completed his Internal Medicine training at the University of Southern California, where he stayed for an additional year as Chief Resident. He has an interest in quality improvement and outcomes research and plans on pursuing a career in non-invasive cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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