

CASE REPORT

Anterior mitral valve aneurysm: a rare sequelae of aortic valve endocarditis

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Summary

In intravenous drug abusers, infective endocarditis usually involves right-sided valves, with *Staphylococcus aureus* being the most common etiologic agent. We present a patient who is an intravenous drug abuser with left-sided (aortic valve) endocarditis caused by *Enterococcus faecalis* who subsequently developed an anterior mitral valve aneurysm, which is an exceedingly rare complication. A systematic literature search was conducted which identified only five reported cases in the literature of mitral valve aneurysmal rupture in the setting of *E. faecalis* endocarditis. Real-time 3D-transesophageal echocardiography was critical in making an accurate diagnosis leading to timely intervention.

Learning objectives:

- Early recognition of a mitral valve aneurysm (MVA) is important because it may rupture and produce catastrophic mitral regurgitation (MR) in an already seriously ill patient requiring emergency surgery, or it may be overlooked at the time of aortic valve replacement (AVR).
- Real-time 3D-transesophageal echocardiography (RT-3DTEE) is much more advanced and accurate than transthoracic echocardiography for the diagnosis and management of MVA.

Background

Mitral valve aneurysm (MVA) is an uncommon condition that can occur as a complication of infective endocarditis of aortic valve or the mitral valve. Rupture of the aneurysm is the most feared complication, which can result in severe mitral regurgitation (MR) causing rapid hemodynamic deterioration especially in heart failure patients (1, 2, 3). A timely diagnosis using Real-time 3D-transesophageal echocardiography (RT-3DTEE) and appropriate treatment such as surgical repair or replacement of the valve can prevent this catastrophic complication (2, 3). We present a case of anterior MVA after aortic valve endocarditis and emphasize the role of RT-3DTEE in the early diagnosis and management of this condition.

Case presentation

A 41-year-old Caucasian male with history of intravenous drug use, chronic obstructive pulmonary disease and colorectal abscesses presented with a 3-week history of worsening shortness of breath. At the time of presentation, the patient endorsed extreme fatigue, night sweats, orthopnea and dyspnea. The patient denied chest pain, cough, nausea, or vomiting. Cardiac examination revealed a diastolic murmur along the left sternal border. Initial workup included computed tomographic angiogram (CTA) of the chest, which showed no evidence of pulmonary emboli. The 2D-transthoracic echocardiogram (TTE) demonstrated a 2.1 cm aortic valve vegetation as well as an abnormal ring-like structure on the mitral valve (Fig. 1A

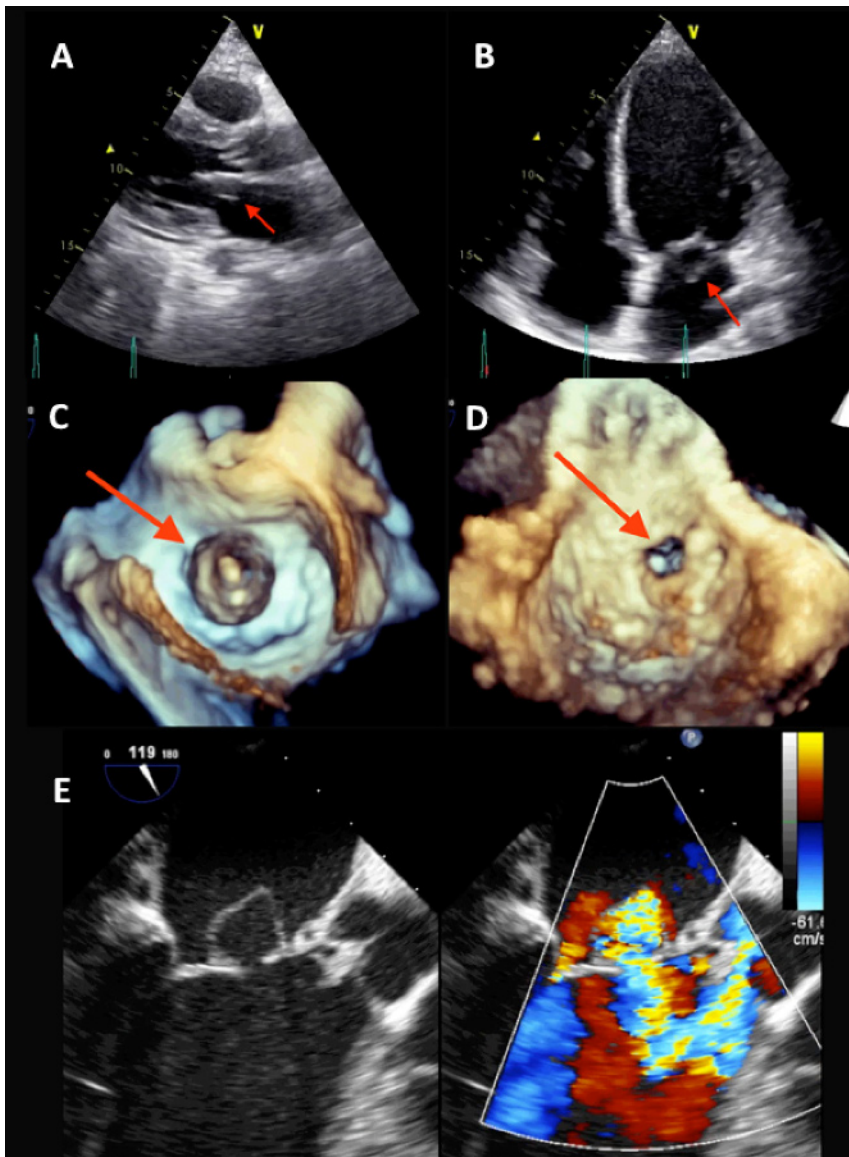


Figure 1

Parasternal long-axis view (A) and apical 4-chamber view (B) on 2D-transthoracic echocardiography showing an abnormal ring-like structure on the mitral valve (red arrow). (C) RT-3DTEE: 3D zoom view from the left atrial side demonstrating an aneurysm (red arrow) involving the A2 scallop of the mitral valve (RT-3DTEE, real-time 3D-transesophageal echocardiography). (D) RT-3DTEE: 3D zoom view from the left ventricular side demonstrating a perforation (red arrow) involving the A2 scallop (RT-3DTEE, real-time 3D-transesophageal echocardiography). (E) Color comparison on color Doppler flow on TEE: The panel on the right shows the aortic regurgitation jet impinging on the undersurface of the anterior mitral valve leaflet (TEE, transesophageal echocardiography).

and B). Left ventricular ejection fraction (LVEF) was 65%. The patient was started on empiric antibiotics, including vancomycin, ceftriaxone and gentamicin immediately after blood cultures were obtained. The patient was transferred to a tertiary center for higher level of care, further evaluation, and management of endocarditis. Upon arrival, an RT-3DTEE was performed, revealing vegetations on all three cusps of the aortic valve, the largest measuring 2.7 cm. There was malcoaptation of the aortic valve cusps with severe aortic regurgitation. 3D zoom view from the left atrial side demonstrated an aneurysm involving the A2 scallop of the mitral valve (Fig. 1C). The view from the left ventricular side revealed a perforation in the A2 scallop (Fig. 1D). Blood cultures returned positive for *Enterococcus*

faecalis bacteremia. The bacteremia was attributed to the use of rectal methamphetamine suppositories complicated by rectal abscesses.

Based on RT-3DTEE findings of severe symptomatic aortic regurgitation and anterior mitral valve aneurysm the patient underwent urgent surgery. Intraoperatively, the patient was found to have a large 2 × 2 cm perforation through A2 of the mitral valve (Fig. 2A). There was a large ballooning piece of tissue seen over the perforation in the A2 scallop of the anterior leaflet, corresponding to the preoperative 3DTEE findings. The excess aneurysmal tissue and leaflet were resected. The aortic valve had several vegetations with ragged edges (Fig. 2B), and there was malcoaptation of the leaflets as demonstrated

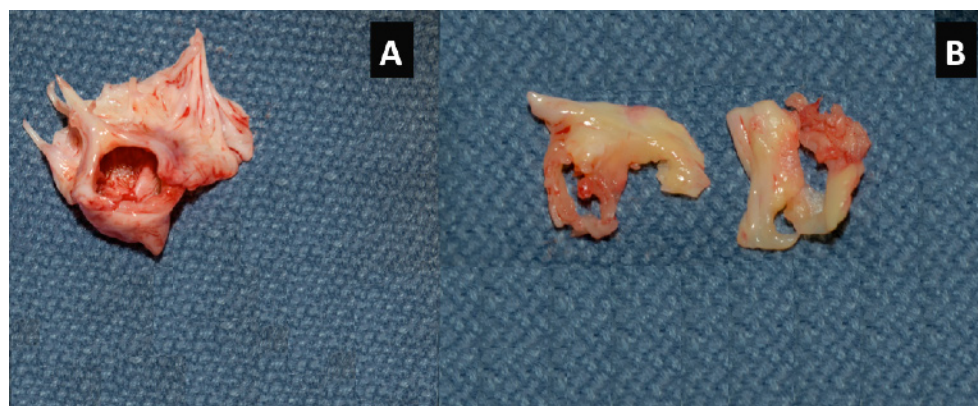


Figure 2

(A) Excised mitral valve showing a large 2 × 2 cm perforation through A2 scallop of the mitral valve. (B) Excised aortic valve with vegetations attached.

on RT-3DTEE. The patient underwent successful aortic and mitral valve replacements. Cultures of the valves also grew *E. faecalis*. The patient was subsequently discharged on ampicillin and streptomycin for 6 weeks and infectious disease follow-up.

Discussion

Our patient developed aortic valve enterococcal endocarditis, most likely due to use of rectal methamphetamine suppositories. The patient subsequently developed colorectal abscesses, requiring a colostomy. Enterococcus is the third leading etiological agent for bacterial endocarditis after viridans streptococci and staphylococci, and accounts for approximately 8% of all cases of bacterial endocarditis (4). The patients at the highest risk for developing enterococcal endocarditis include elderly men subjected to multiple genitourinary procedures, young women with postpartum and genitourinary infections, and intravenous drug users (5). These patients commonly present with fever, night sweats, weight loss, malaise, heart murmurs and symptoms due to cardiac failure.

MVA is a rare complication. To the best of our knowledge, only 10 cases of MVA in enterococcal endocarditis have been reported in the literature (Tables 1, 2 and 3). A further systematic literature search was conducted using Medline (via PubMed), Embase, Scopus and Google Scholar, which identified only five reported cases from *E. faecalis* endocarditis (Table 1). The typical occurrence is in the presence of aortic valve endocarditis, rheumatic disease and other disorders causing connective tissue degeneration (3, 6, 7). Rarely, MVA has been reported in noninfectious etiologies like connective

tissue disorders such as Marfan syndrome, Ehlers–Danlos syndrome, pseudoxanthoma elasticum and some cases of mitral valve prolapse (8, 9, 10, 11).

MVA is a rare condition with reported incidence of 0.29% on 4500 TEE examinations (12). On TTE it looks like a saccular bulge of the mitral leaflet protruding toward the left atrium with systolic expansion and diastolic collapse. The diastolic expansion may occur with AR or after rupture of the MVA (13). Anterior MVA is more commonly observed than the posterior MVA (13, 14). The development of MVA is likely due to the infected aortic regurgitant jets striking the ventricular surface of the anterior mitral leaflet (Fig. 1E) causing physical trauma and possible occult mitral leaflet infection. This is manifested by valvulitis and the formation of sac-like outpouchings due to formation of scar tissue and granulation tissue on microscopic examination. The extension of infection to the mitral–aortic intervalvular fibrosa results in abscess or aneurysm formation (1, 2, 10). However, the posterior MVA occurs as a result of weakness of the mitral valve secondary to myxomatous degeneration and latent infective endocarditis (15). The MVA is of variable shape and size and can easily be confused with other mitral valve masses. According to Gular and coworkers, these aneurysms range in size from 5 to 12 mm in diameter during ventricular systole. The differential diagnosis of MVA includes mitral valve prolapse, myxomatous degeneration of the mitral valve, flail mitral leaflet, papillary fibroelastoma, myxoma involving the mitral valve, and mitral valve blood cysts without endothelialization and mitral valve diverticulum (13). The potential complications of MVA include endocarditis, thromboembolization and rupture of the aneurysm or perforation of the valve leaflet leading to acute, severe mitral regurgitation and pulmonary edema

Table 1 Cases reported of MVA in the setting of *Enterococcus faecalis* endocarditis.

| Case No. | Study (year) | Age (years) and sex | Predisposition | Presentation | Cardiac examination findings | Causative organism | MV cusps involved | Echocardiography findings | MVA perforation | AV replacement | MV replacement |
|----------|----------------------------------|---------------------|--|--------------------------------|---|------------------------------|-------------------|---|-----------------|----------------|----------------|
| 1 | Kholeif <i>et al.</i> (2002) | 33 M | Nephrotic syndrome, Steroid use | Fever | PSM at apex and diastolic murmur at aortic area | <i>Enterococcus faecalis</i> | AMV | TEE: Thickened MV with an echo-dense mass, 3 large vegetations on AV, moderate MR and severe AR | 0 | + | + |
| 2 | Körber <i>et al.</i> (2001) | 65 M | AR | Pulmonary edema | Grade IV PSM over mitral area | <i>Enterococcus faecalis</i> | AMV | TEE: Aneurysm on AMV, severe MR | + | + | + |
| 3 | Rachko <i>et al.</i> (2001) | 63 M | UTI | Chest pain, dyspnea, dizziness | Unknown | <i>Enterococcus faecalis</i> | AMV | TTE: Thickened AV and MV leaflets TEE: AV vegetation, saccular structure with a narrow neck attached to AMV, severe MR and AR | 0 | + | 0 |
| 4 | Seratnahaei <i>et al.</i> (2015) | 29 M | Enterocutaneous fistulas | Fever and delirium | Unknown | <i>Enterococcus faecalis</i> | AMV | TEE: MV vegetation and aneurysm perforation | + | + | + |
| 5 | Seratnahaei <i>et al.</i> (2015) | 61 M | <i>Clostridium difficile</i> infection | Fever | Unknown | <i>Enterococcus faecalis</i> | AMV | TEE: MV vegetation and AMV aneurysm perforation | + | 0 | 0 |

Positive, +; Negative, 0; AMV, anterior mitral valve; MV, mitral valve; PMV, posterior mitral valve; AS, aortic stenosis; AR, aortic regurgitation; MR, mitral regurgitation; TTE, transthoracic echocardiography; TEE, transesophageal echocardiography.

Table 2 Cases reported of MVA in the setting of *Enterococcus faecium* endocarditis.

| Case No. | Study (year) | Age (years) and sex | Predisposition | Presentation | Cardiac examination findings | Causative organism | MV cusps involved | Echocardiography findings | MVA perforation | AV replacement | MV replacement |
|----------|----------------------------------|---------------------|--------------------|------------------------|---|-----------------------------|-------------------|---|-----------------|----------------|----------------|
| 1 | Domínguez <i>et al.</i> (1998) | 34 | Ulcerative colitis | Fever | Diastolic murmur and ESM at aortic area, S3 | <i>Enterococcus faecium</i> | AMV | TTE: AMV aneurysm, Doppler: severe AR and moderate MR | + | + | 0 |
| 2 | Hotchi <i>et al.</i> (2011) | 77 F | – | Incidental | Unknown | <i>Enterococcus faecium</i> | PMV | TTE and TEE: severe MR and AR, cystic mobile lesion on the PMV | 0 | + | + |
| 3 | Pederzollia <i>et al.</i> (2009) | 67 M | Aortic stenosis | Chest pain and dyspnea | PSM at apex and grade-III diastolic murmur at LPB | <i>Enterococcus faecium</i> | PMV | 3DTEE: severe MR due to large perforated aneurysm of the posterior leaflet (Scalloped P3) | + | + | + |

Positive, +; Negative, 0; AMV, anterior mitral valve; MV, mitral valve; PMV, posterior mitral valve; AS, aortic stenosis; AR, aortic regurgitation; MR, mitral regurgitation; TTE, transthoracic echocardiography; TEE, transesophageal echocardiograph.

Table 3 Cases reported of MVA in the setting of enterococcal endocarditis (unspecified).

| Case No. | Study (year) | Age (years) and sex | Predisposition | Presentation | Cardiac examination findings | Causative organism | MV cusps involved | Echocardiography findings | MVA perforation | AV replacement | MV replacement |
|----------|------------------------------------|---------------------|------------------------|--------------|------------------------------|--------------------|-------------------|--|-----------------|----------------|----------------|
| 1 | Isidre Vilaco <i>et al.</i> (1999) | 55 F | Heart failure | Dyspnea | Unknown | Enterococcus | AMV | TEE showing MVA of 2×3 mm with severe MR and severe AR | + | + | + |
| 2 | Isidre Vilaco <i>et al.</i> (1999) | 55 F | Heart failure embolism | Dyspnea | Unknown | Enterococcus | AMV | TEE showing MVA of 12×16 mm with severe MR and moderate AR | + | + | + |

Positive, +; Negative, 0; AMV, anterior mitral valve; MV, mitral valve; PMV, posterior mitral valve; AS, aortic stenosis; AR, aortic regurgitation; MR, mitral regurgitation; TTE, transthoracic echocardiography; TEE, transesophageal echocardiography.

(10, 16). Vilacosta and coworkers studied the natural course of these aneurysms with serial echocardiographic follow-up and concluded that they undergo progressive expansion and subsequent rupture or perforation (12). The vegetation or thrombus formation may occur within the aneurysm leading to thromboembolism and spread of infection (11).

Transthoracic echocardiography can demonstrate MVA as a localized saccular bulge of the mitral valve leaflet toward the left atrium, which persists throughout the cardiac cycle. However, it can be challenging to localize the exact site and size of aneurysmal rupture because of the inherent limitations of 2D-transthoracic echocardiography (16). Therefore, the presence of mitral aneurysm may be missed in important clinical situations such as critically ill patients with aortic valve endocarditis or during the preoperative cardiac assessment of mitral valve prior to aortic valve replacement in setting of endocarditis (3, 6, 7, 17). Hence, RT-3DTEE, which is superior to TTE for the definitive diagnosis and precise determination of MVA, should be employed to guide management decisions in such circumstances. This case clearly demonstrates that the use of RT-3DTEE was critical in making an accurate diagnosis and planning the appropriate surgical approach with a successful outcome in our patient with this rare presentation (16).

A conservative approach for small, uncomplicated aneurysms is a reasonable option with close follow-up; but surgical options are utilized in cases with large unruptured aneurysms or in the setting of perforation or rupture of the aneurysm with or without significant MR (18). Mitral valve repair in the setting of infective endocarditis has been shown to have good clinical in-hospital and long-term results as compared to MV replacement (19, 20). The effective utilization of 3DTEE is invaluable for the success of surgical repair in these settings (20). The accurate determination of the size of the perforation and preoperative 3D pictures of the MVA utilizing TEE is tremendously helpful for operative planning. The RT-3DTEE images and operative findings coincided in 92% of cases in a study conducted by Kanzaki and coworkers. The diagnostic capability of 3D-TEE depends on the leaflet segment or scallop with 67–100% and 100% sensitivity for detecting lesions at anterior and posterior leaflet, respectively; and specificities of 78–100% and 100% at anterior and posterior leaflet, respectively (21).

Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this case report.

Funding

This research did not receive any specific grant from any funding agency in the public, commercial or not-for-profit sector.

Patient consent

Written informed consent was obtained from the patient for publication of the submitted article and accompanying images.

Author contribution statement

R J: Conception and design, and final approval of manuscript. M U K: Conception and design, and drafting of manuscript. I B R: Conception and design, and drafting of manuscript. C S: Conception and design, and approval of manuscript.

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Received in final form 7 March 2016

Accepted 10 March 2016

Accepted Preprint published online 10 March 2016