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LETTER TO THE EDITOR

Sequential spontaneous severe aortic and mitral regurgitation

KEYWORDS
Aortic valve replacement;
Mitral valve replacement;
Rheumatoid process;
Polymyalgia rheumatica

Acute severe valve regurgitation has been described in both aortic and mitral valves. In the aortic valve, regurgitation is most commonly a result of infection, aortic dissection, aortic aneurysms or trauma, whereas for the mitral valve, acute regurgitation is generally a result of infection, chordal rupture or ischemia. Rheumatologic disorders affecting cardiac valves, although rare, have been reported to cause both spontaneous aortic and mitral insufficiency. This report, however, is a rare case describing sequential inflammatory damage to both valves in the same patient, requiring two surgical sessions 13 months apart.

A 58 year old female patient was referred for aortic valve replacement. She was admitted with an episode of acute onset dyspnea associated with chest discomfort. Echocardiography revealed severe aortic regurgitation with a prolapsing non-coronary cusp (NCC). Left ventricular function was moderately impaired, and the left ventricle was mildly dilated. Coronary angiography showed normal coronary anatomy. She had a history of chronic obstructive pulmonary disease, peripheral vascular disease, smoking and polymyalgia rheumatica for which she was treated with steroids. She had no clinical or serological evidence of rheumatoid arthritis.

She was operated on an urgent basis, and she underwent an AVR using a mechanical valve prosthesis. The most striking intraoperative finding was the complete prolapse of the NCC (Figure 1a). The right and left coronary cusps appeared normal, while there was no evidence of infection, aortic aneurysm or dilatation. She recovered well from the operation and was discharged home.

Histopathological findings from the aortic valve specimen revealed endothelialitis with inflammatory changes (Figure 2a). These findings were related to her history of polymyalgia rheumatica. All tissue cultures were negative, and the Gram stain on the tissue did not identify any organisms.

The patient presented 13 months later to her family doctor describing an acute onset 'humming noise' from her left chest, which became louder upon exertion. Interestingly, the murmur was loud, systolic and audible even without a stethoscope. She did not complain of any other symptoms at that time. She underwent transthoracic echocardiography that revealed severe mitral regurgitation with prolapse of the anterior mitral valve leaflet, some calcification on the posterior mitral valve leaflet and evidence of chordal rupture (Figure 1b,c). The aortic valve prosthesis was well-seated and functioning as expected. There was no evidence of endocarditis or ischemic event. As she was initially asymptomatic, she was initially treated conservatively. After 3 months, however, she developed progressively worsening symptoms indicating the need for reoperation.

She underwent a redo-sternotomy mitral valve replacement with a mechanical prosthesis. The mitral valve macroscopically appeared to have a mild degree of myxomatous degeneration. The anterior leaflet was flail with the presence of ruptured chords. The posterior mitral annulus was mildly calcified. The valve was replaced by using a leaflet-sparing technique.

The histopathology report of the mitral valve specimen showed evidence of inflammation and endothelialitis. Though a definite rheumatoid nodule was not identified, damaged connective tissue was present in a pattern consistent with damage from a rheumatologic process (Figure 2b). All microbiology investigations were negative.

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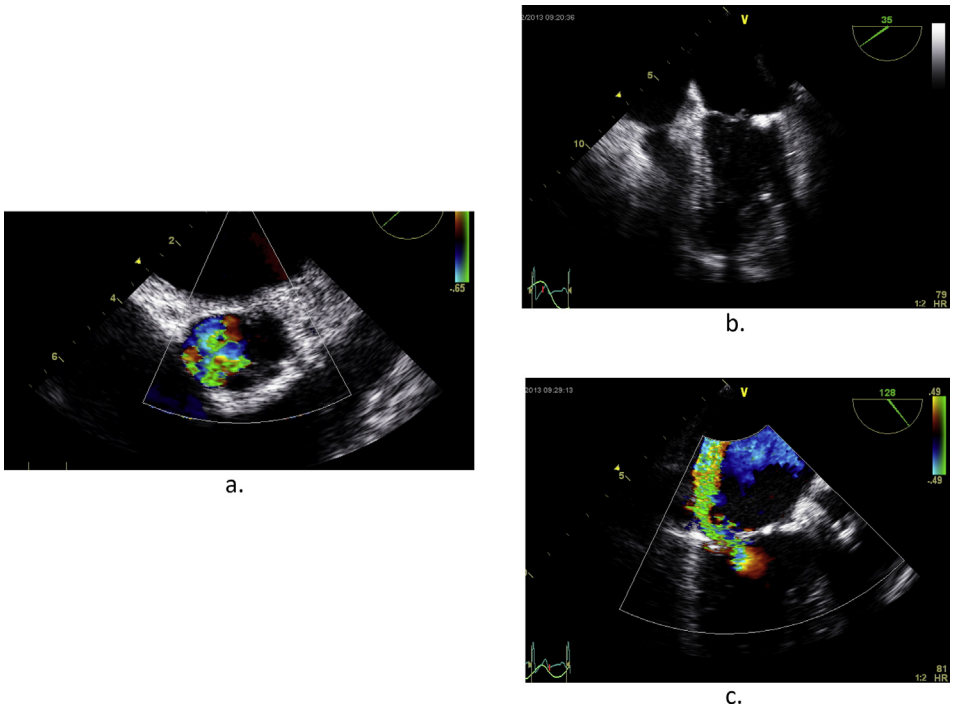

Figure 1 On-table preoperative transesophageal echocardiogram showing: a. the aortic valve in the short-axis view demonstrating the severe AI originating from the NCC. b. the prolapsing anterior mitral valve leaflet. c. the resulting severe posteriorly directed jet on color Doppler flow

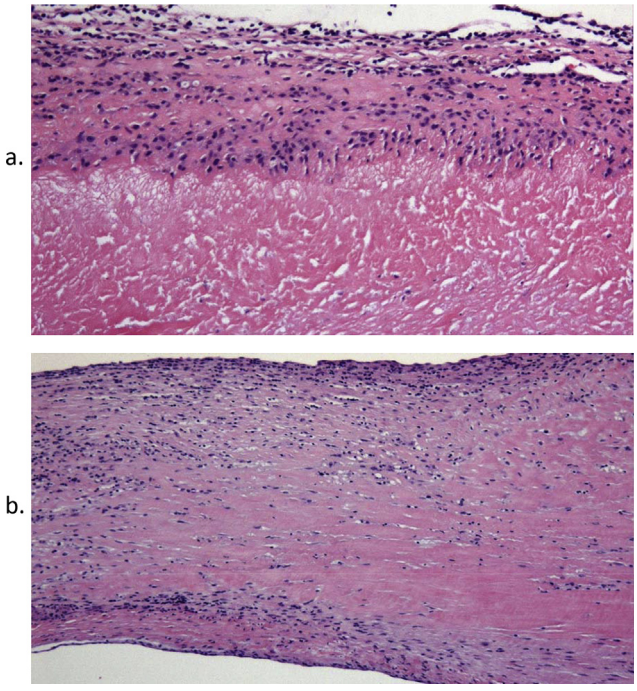

Figure 2 Histopathological specimens showing: a. Aortic valve specimen with endothelialitis along the surface. The underlying tissue contains a zone of necrotic tissue surrounded by a cuff of palisading mononuclear cells. (H&E x 100). b. Mitral valve specimen with endothelialitis along the surface. The underlying tissue contains inflammation and damaged collagen (H&E x 50).

The patient was discharged home 6 days after her second operation. The pre-discharge transthoracic echocardiography revealed improved biventricular function, with both the mitral and aortic prostheses functioning as normal. She was well on follow-up six weeks after her second operation without any symptomatology or significant clinical findings.

Spontaneous aortic valve regurgitation caused by acute rupture of one of the leaflets has been described in the literature. This regurgitation was related to myxomatous changes of the aortic valve in the absence of infection.[1,2] Similarly, aortic regurgitation necessitating aortic valve replacement has been described in patients with rheumatoid arthritis. In this group of patients, the pathophysiology ranged from aortitis with dilatation to inflammatory degeneration and prolapse of the valve leaflet.[3,4]

Double and triple valve involvement have been described in very limited case reports in the literature, and in all of them, the patients had a history of rheumatoid arthritis.[5,6] Interestingly, there is no published report that associates polymyalgia rheumatica with inflammatory cardiac valve involvement. This lack of association may be because heart valve manifestation is rare in inflammatory diseases. In our case, despite the fact that our patient had been diagnosed with polymyalgia rheumatica, one cannot exclude that the patient could also be suffering from a subclinical type of rheumatoid arthritis. In our case, the 'sequential' presentation of the two valves involved within 13 months appears to be extremely rare.

To conclude, we present a unique case of sequential double valve acute insufficiency caused by inflammation

possibly related to a rheumatoid process on a patient suffering from polymyalgia rheumatica.

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