



## Mitral-Aortic Intervalvular Fibrosa Involvement by Takayasu' Arteritis

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### ABSTRACT

Takayasu's arteritis is an inflammatory disease with a variety of manifestations, such as cardiac involvement. We describe a 52-year-old woman with clinical and echocardiographic manifestations mimicking infectious endocarditis, such as periaortic and mitral-aortic intervalvular fibrosa abscess with extension to the anterior mitral leaflet. However, no infective tissue was discovered intraoperatively. Pathological evaluation demonstrated Takayasu's arteritis. To the best of our knowledge, Takayasu's arteritis can involve mitral-aortic intervalvular fibrosa and imitate infectious endocarditis.

### ► Implication for health policy/practice/research/medical education:

This case expresses rare presentation of takayasu's arteritis that simulated infectious endocarditis of mitral and aortic valve.

### 1. Introduction

Takayasu's Arteritis (TA) is an idiopathic inflammatory fibrosing arteritis affecting predominantly the aorta and its main side branches, most commonly in young women (1). Aortic regurgitation is a relatively common important complication observed in these patients, resulting from the inflammation of the aortic root and valve (1, 2). Increased aortic wall thickness is a common feature of TA in echocardiography (3). TA can involve mitral valve (4) and mitral- aortic intervalvular fibrosa (5).

Low operative mortality and favorable long-term outcomes may justify aortic valve replacement in the patients with significant aortic regurgitation secondary to TA (6). However, late enlargement of the ascending aorta and valve detachment after aortic valve replacement can be due to active inflammation (7).

Here, we describe a patient with TA accompanied by severe aortic regurgitation, resembling mitral-aortic intervalvular fibrosa and periaortic abscess in echocardiography.

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### 2. Case Presentation

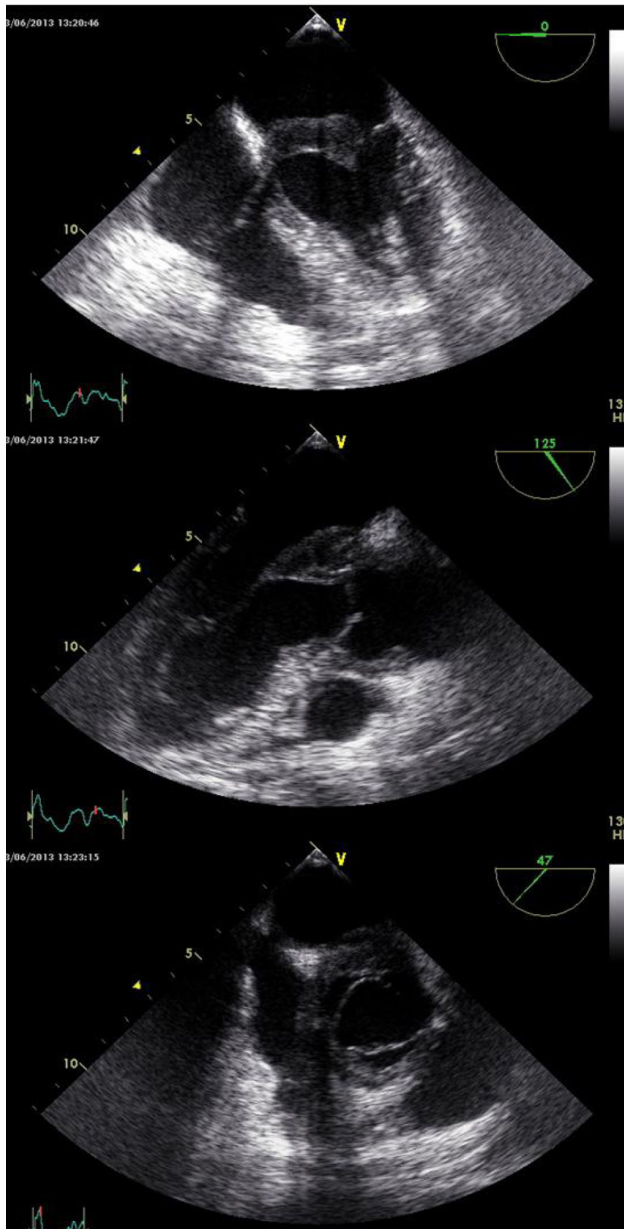
A 52-year-old woman presenting with fever, weakness, nausea, headache, and progressive dyspnea on exertion of 3 months duration referred to our hospital with the diagnosis of aortic valve endocarditis. Infertility and chronic gastritis were the highlights of her past medical history.

In physical examination, blood pressure was 130/60 mmHg in the left arm with no difference from the other arm, showing a widened pulse pressure with a low diastolic pressure. Upper and lower limb pulses were full and symmetrical. Diastolic murmur was heard at the left sternal border. There was no audible bruit on the carotid arteries and no tenderness on the temporal arteries. There was no history of claudication in the limbs.

First-degree atrioventricular block with incomplete right bundle branch block was found in the patient's electrocardiogram. Moreover, notable laboratory examinations were mildly elevated white blood cell count of 10210 / $\mu$ L, mild anemia (Hb = 10.1 mg/dL), erythrocyte sedimentation rate of 90 mm/h, and C-Reactive Protein (CRP) of 6.06 mg/dL.

Transthoracic and transesophageal echocardiographic

examinations revealed normal left ventricular volume and systolic function (ejection fraction = 55%) with severe central aortic regurgitation and increased anterior and posterior aortic root thickness (12 and 16 mm, respectively). The thickened aortic root extended to the mitral-aortic intervalvular fibrosa and two thirds of the basal part of the anterior mitral leaflet (15 mm) and resulted in the thickening of the anterior mitral valve. This was in favor of aortic root and mitral-aortic intervalvular fibrosa abscess formation. There was also extension to the anterior mitral valve (Figure 1) and moderate posterolaterally-directed mitral regurgitation, without any vegetation on the mitral or aortic valves. Moreover, the ascending aorta was normal in size.



**Figure 1.** Anterior Mitral Valve Thickening in Transesophageal Echocardiography (Four-Chamber View) (Upper), Anterior Mitral Valve Thickening and Aortic Root Wall Thickening in Transesophageal Echocardiography (Aortic Valve Long-Axis View) (Middle), and Aortic Root Wall Thickening in Transesophageal Echocardiography (Aortic Valve Short-Axis View) (Lower)

Intraoperatively, the remarkable findings were thickened

ascending aorta and aortic arch, with extension to the mitral-aortic intervalvular fibrosa and anterior mitral valve leaflet, without vegetation or infectious tissue. Accordingly, aortic valve replacement with a mechanical bileaflet prosthetic valve was performed without mitral valve repair or replacement in the hope of resolving the mitral regurgitation with anti-inflammatory drugs.

Biopsy specimens of the aortic wall disclosed medial cystic degeneration, elastic fiber fragmentation, adventitial fibrosis with granulomatous-like inflammation, and lymphocyte and macrophage infiltration. In addition, giant cell granulomatous reaction with inflammatory reaction was found in the mitral valve, which was in favor of TA because of adventitial involvement. There was no evidence of bacterial infection. Therefore, the patient was treated with Methylprednisolone pulses followed by oral Prednisolon. Transthoracic echocardiography 2 weeks after the surgery showed new moderate paravalvular leakage.

### 3. Discussion

The criteria for diagnosis of TA, set by the American College of Rheumatology, consist of 1- age at disease onset < 40 years, 2- claudication of the extremities, 3- decreased brachial artery pulse, 4- systolic blood pressure difference between the two arms > 10 mmHg, 5- bruit over the subclavian arteries or aorta, and 6- arteriogram abnormality. The presence of three or more criteria yields a sensitivity of 90.5% and a specificity of 97.8% (8). Our patient exhibited none of these criteria. She, however, demonstrated only inflammatory signs (as a key manifestation) and clinically possible endocarditis. Periaortic, mitral-aortic intervalvular fibrosa, and thickening of almost the entire anterior mitral leaflet led to the false notion of infectious endocarditis. Indeed, these features rendered a correct diagnosis prior to the surgery extremely difficult. Be that as it may, absence of vegetation and perforation of the leaflets were the salient points that were crucially neglected.

Circumferential thickening of the thoracic aorta, a common transesophageal echocardiographic finding in TA, affects a significant proportion of the segments (3). Mitral valve involvement in the form of mitral regurgitation (4), mitral stenosis (9), and mitral annular calcification (10) have been reported, as well. Additionally, there are reports of the pseudoaneurysm of the mitral-aortic intervalvular fibrosa (5). Nonetheless, the existing literature contains no reports on mitral-aortic intervalvular fibrosa thickening with extension to two thirds of the basal part of the anterior mitral leaflet. Paravalvular leakage after a short period appeared in our patient, but it was predictable (7).

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### Authors' Contribution

Study concept and design, Collection of data, Drafting of the manuscript, Critical revision of the manuscript for important intellectual content, Statistical analysis, Administrative, technical, and material support: all authors; Analysis and interpretation of data, Study supervision: Tahereh Davarpasand, Ali Hosseinsabet

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