

Coexistence of Aortic Pseudocoarctation and Mitral Valve Perforation in a Patient With Takayasu's Arteritis: Coincidence or Association?

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ABSTRACT

Takayasu's arteritis (TA) is a rare idiopathic inflammatory condition that predominantly involves the aorta and its branches causing narrowing and aneurysms in vessels. In this case report, we present TA with a rare coexistence of aortic pseudocoarctation and anterior mitral leaflet perforation with moderate regurgitation.

Keywords: Aortic pseudocoarctation; computed tomography; mitral valve perforation; Takayasu's arteritis; two- and three-dimensional transesophageal echocardiography.

Takayasu's arteritis (TA) is a rare, systemic, inflammatory large-vessel vasculitis of unknown etiology that most commonly affects females of childbearing age and causes narrowing and aneurysmal formation of the aorta as well as aortic regurgitation.¹⁻³ However, concomitant stenosis and dilatation of aorta, and anterior mitral leaflet perforation leading to moderate regurgitation is very rare with TA. In this article, we report a case of TA, which is associated or coincidental with other comorbidities.

CASE REPORT

A 26-year-old female patient with eight-year history of TA had undergone Bentall procedure involving composite replacements of aortic root with 30 mm Dacron tube graft and St. Jude, No. 23 mechanical aortic valve (St. Jude Medical

Drive, St. Paul, Minnesota, USA) due to severe aortic aneurysm and valve regurgitation seven years ago and presented with chest pain and labile hypertension. She was treated with combined immunosuppressive therapy of prednisolone and azathioprine for five years. Additionally, the patient was on warfarin treatment with a target international normalized ratio between 2-4 for prosthetic aortic valve replacement. On physical examination, she had rhythmic heart beats, 3/6 systolic murmur on the fifth left intercostal space. Her blood pressure on the right arm was 150/96 mmHg and there was no pressure difference between her right arm and legs. Routine blood tests showed that inflammatory markers and white blood cell counts were within normal limits and two blood culture sets taken on admission were negative. Electrocardiography revealed a normal sinus rhythm with a ventricular rate of 75 beats/minute. Comprehensive transthoracic

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Figure 1. Two-dimensional transesophageal echocardiography shows 2 mm-leaflet perforation with moderate regurgitation on medial side of native mitral valve (plus). LA: Left atrium; LV: Left ventricle.

and real-time two- and three-dimensional transesophageal echocardiography showed 2 mm-anterior leaflet perforation with moderate regurgitation on medial side of native mitral valve as well as normal left ventricular function and wall motion, and functionally normal aortic root replacement with mechanical composite aortic valve (Figure 1). There was no annular calcification, and no valvular vegetations were seen. Three-dimensional thoracoabdominal computed tomography revealed aneurysm and coarctation of aorta with diffuse calcifications on the descending aorta (Figure 2). Conventional aortography was performed using a pigtail catheter which revealed an aortic gradient of 15-20 mmHg. Since there was no significant hemodynamic gradient and moderately mitral valve regurgitation, medical treatment was advised. Written informed consent was obtained from the patient.

DISCUSSION

Takayasu's arteritis is characterized by stenocclusive disease of the aorta and its main branches due to idiopathic chronic vasculitis. The pathogenesis of this disease remains unclear.^{4,5} The clinical course of TA is thought to progress through nonspecific systemic symptoms such as fatigue, low-grade fever, and weight loss, and depending on the vessels involved, through a wide variety of vascular or ischemic manifestations.^{6,7} Aortic coarctation and TA have

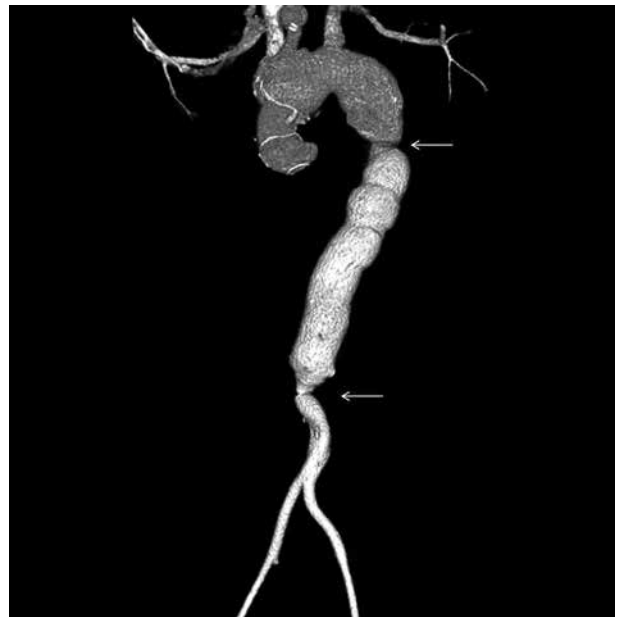


Figure 2. Three-dimensional thoracoabdominal computed tomography reveals aneurysm and coarctation of aorta (arrows) and also aortic graft with mechanical prosthetic aortic valve.

clinical similarities which should be kept in mind during diagnostic work up.⁸ The coexistence of aortic pseudocoarctation and TA has been reported occasionally secondary to inflammatory mechanisms.⁹ Although endovascular intervention of aortic stenosis is less invasive compared to surgical options, resulting in fewer procedural complications and lower mortality in patients with severe aortic gradients, surgical repair seems to be the first choice in the treatment of aortic coarctation when associated with aneurysmal degeneration, unfavorable anatomy, including severe occlusion of the aorta or extension to the aortic bifurcation and severe hypertension.¹⁰

We also present here an incidentally detected mitral valve perforation (MVP) in an adult patient with undetermined cause. It is usually caused by infective endocarditis.¹¹ Valve perforation can occur by either primary vegetation destroying valve tissue, secondary mitral valve endocarditis caused by an endocarditic aortic valve jet lesion, or direct contact of the mitral leaflet with mobile vegetation on the aortic valve.^{12,13} However, congenital or iatrogenic conditions, including previous aortic graft with mechanical prosthetic aortic valve operations may lead to mitral leaflet

perforation. In our patient, we were not able to define the underlying mechanism for mitral perforation. Spontaneous mitral leaflet perforation is very rare, with a paucity of published data in the literature. Additionally, simultaneous presence of TA and aortic pseudocoarctation and MVP is unexpected and raises the possibility of either causal or coincidental association between these conditions.

In conclusion, inflammatory manifestations of TA are various and poorly understood. The coexistence of TA and aortic pseudocoarctation and MVP is very rare but probably not coincidental. TA may cause aortic narrowing and aneurysm as well as MVP by stimulating inflammatory processes.

Declaration of conflicting interests

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