A Case of Aortic Regurgitation and Hypotension

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Int J Angiol 2013;22:263–266.

Abstract

Keywords

- ▶ Takayasu arteritis
- ▶ aortic regurgitation
- ▶ vasculitis
- ▶ hypotension

Takayasu arteritis is a chronic inflammatory large vessel vasculitis affecting the aorta and its main branches. It can present in various forms, and thereby may lead to a delay in diagnosis. We present a patient with Takayasu arteritis, diagnosed at our center when she presented with severe aortic regurgitation. We also review the current knowledge base with respect to Takayasu arteritis.

Background

Takayasu arteritis primarily affects young women. It is a chronic, inflammatory disease affecting large vessels and its main branches, but its effects can manifest systematically in the form of various ischemic symptoms.

Case Presentation

We present a 26-year-old Indian lady who came to us for a second opinion regarding possible aortic valve replacement.

She had presented to another hospital and was diagnosed with symptomatic anemia to account for her giddiness, malaise, and nonspecific upper limb pain. These symptoms had been unremitting for months. Although her symptoms initially improved after blood transfusions, they recurred a few weeks later. A loud cardiac murmur was then heard and she was diagnosed with aortic regurgitation (AR) and referred for aortic valve replacement. However, she was keen for a second opinion and further evaluation and therefore presented to our center.

On physical examination, she did not have any stigmata of infective endocarditis. Her physical appearance was not suggestive of any particular congenital syndrome. Significantly, her radial and brachial pulses were impalpable. There was a marked difference in the blood pressure (BP) between her upper and lower limbs—upper limb BP was 75/50 as compared with lower limb BP of 150/90. On auscultation of her precordium, there was a diastolic murmur as well as a loud bruit throughout. She was clinically not in heart failure.

Laboratory investigations revealed microcytic, hypochromic anemia with a hemoglobin of 10.8 g/dL. Erythrocyte sedimentation rate (ESR) was elevated at > 140 mm/h. There

was a mildly raised serum alkaline phosphatase of 105 U/L and a low serum albumin of 33 g/L. The rest of the routine blood investigations were within normal limits. Blood cultures were negative.

Chest radiograph demonstrated a prominent ascending aorta and aortic arch silhouette. The cardiothoracic ratio was normal.

Transthoracic echocardiography revealed severe AR probably secondary to a dilated aortic root of 3.3 cm (aortic root index calculated to be 4.98, according to the Dubois formula) (►Fig. 1). The left ventricular ejection fraction was preserved at 50%.

We proceeded to perform a computed tomography aortogram that showed arterial wall thickening of the aorta and aortic arch branches, long segment stenoses of the brachiocephalic artery, bilateral subclavian arteries, right common carotid artery from the origin to the mid common carotid artery, the entire left common carotid artery, and bilateral vertebral arteries (►Fig. 2). The cerebral circulation was mainly dependent on the left vertebral artery. There was also a saccular aneurysm noted arising from the ascending aorta.

Magnetic resonance imaging and angiography (MRA) of the brain showed diffuse prominent enhancement of bilateral cerebral cortical veins, which may have indicated a degree of delayed transit in cerebral perfusion. There was no significant intracranial stenosis detected on MRA of the circle of Willis. Decreased flow signal and caliber of both internal carotid arteries were noted, worse on the left, secondary to the known severe proximal common carotid stenoses.

Doppler ultrasound of renal arteries did not show any significant renal artery stenosis.

Fig. 1 Dilated aortic root on transthoracic echocardiogram.

From the imaging studies as well as the clinical picture, we diagnosed Takayasu vasculitis, as she fulfilled five out of six of the American College of Rheumatology criteria for diagnosis.

She was started on intravenous hydrocortisone 100 mg every 6 hours. Before initiation of steroids, she was screened for hepatitis and HIV. Interestingly, her hepatitis C antibody screen came back as positive. However, confirmatory RNA viral load studies were negative.

The intravenous steroids were converted to oral prednisolone after 2 days. Although she complained of blurring of vision, ophthalmology review did not identify any significant findings such as retinal hemorrhage/ischemia.

ESR was repeated after 5 days of initiating steroids and it had decreased to 62 mm/h. Clinically, she made a rapid recovery and had essentially no symptoms at the time of

Fig. 2 Reconstructed aorta from computed tomographic imaging.

discharge. She was discharged with oral steroids and the plan was to continue outpatient review with cardiology and rheumatology.

Discussion

Takayasu arteritis was first reported in 1908 by Mikito Takayasu, a Japanese ophthalmologist, as a case of “peculiar changes in the central retinal vessels.” Although it often presents with asymmetrical pulses, Takayasu arteritis is commonly known as the “pulseless disease.” It is essentially a panarteritis affecting the large vessels.

It is now no longer thought to be a disease affecting only young Asian women as more new cases are identified worldwide. The symptoms that patients present with may vary widely, therefore leading to a delayed diagnosis and thereafter treatment.

The lesions are usually on the aorta and its main branches such as the common carotid, vertebral, brachiocephalic, carotid, and vertebral arteries. All these arteries were affected in our patient and we believe that her initial symptoms were most likely due to a lack of cerebral perfusion and upper limb claudication.

AR occurs among 13 to 25% of patients with Takayasu arteritis and it is of prognostic significance. It was first described in 1954 by Jervell and is thought to result from annular dilatation due to dilatation of the ascending aorta.¹ The AR jet causes the aortic valve leaflets to undergo fibrous thickening and results in worsening of the insufficiency.

Management of the aortic valve should be mainly medical unless the valvular insufficiency has caused left ventricular failure. The surgery itself is fraught with higher complication rates—the most important complication of which is subsequent detachment of the prosthetic valve. In a case series of 90 patients who underwent aortic valve replacement for Takayasu arteritis, Matsuura et al reported overall hospital mortality of 5.6%, and survival rates at 5, 10, and 15 years were 87.8, 78.6, and 76.1%, respectively. In the same group, 13.3% had neurological complications such as cerebral infarction, cerebral hemorrhage, and spinal infarction.²

In addition to weakening of the vascular wall causing dilatations, the panarteritis can also cause thickening of the adventitia, media, and intima. This leads to hypertension, pulmonary hypertension, and aortic aneurysm.^{3,4} There can also be concomitant cardiomyopathy, myocarditis, and pericarditis possibly as part of the inflammatory process. The retinopathy that Takayasu first described is seen in 25% of patients and is usually associated with carotid artery involvement.⁵ Our patient has an 8-mm saccular aneurysm of the ascending aorta; such saccular and fusiform aneurysms are found in 31.9% of patients.⁶ Sueyoshi et al suggest that the growth of aneurysms in Takayasu arteritis is related to BP rather than location.⁷ In the same study, it was noted that the growth of aortic aneurysms in Takayasu arteritis at 0.03 cm/y is slower than that seen in atherosclerosis (0.34 cm/y). This is presumably because of more severe aortic wall scarring.

Matsubara et al reported coronary artery involvement in 9 to 10% of the patients that they studied.⁸ As patients do not

commonly complain of angina, coronary artery involvement is usually diagnosed at autopsy. Coronary lesions typically involve the ostia or the proximal segments of the arteries and can be stenotic, occlusive, or aneurysmal with areas of arterial narrowing.

Although symptom relief of claudication is important, recent studies examining the outcomes of percutaneous transluminal angioplasty (PTA) have not been promising. In a reported study, the initial success rate was only 56%, and complications such as restenosis, thrombosis, hemorrhage, infection, or aneurysm formation were reported in 45% of the treated patients.⁹ Arterial bypass and reconstruction procedures in Takayasu arteritis have been shown to be better than PTA although they have their own limitations. In the same case series, 24% of operated patients had restenosis and 4% developed bypass graft thrombosis.

Hepatitis C has been associated with cryoglobulinemic vasculitides and there is now a suggestion of an association between hepatitis C and Takayasu arteritis.¹⁰ However, our patient's confirmatory test was negative and this suggests that Takayasu arteritis resulted in a false-positive result.

The impact of Takayasu arteritis needs to be taken in the context of the afflicted patient population. Patients are typically young and as the nature of the disease is chronic and relapsing, this leads to substantial morbidity and ultimately mortality. Even in patients who are thought to be clinically quiescent, surgical specimens have shown active vasculitis in 44%.⁹ Mortality rate is reported to be 15% by Lupi-Herrera et al⁴ in the 19 years follow-up.

In another study from India, consisting of a cohort of 88 patients, the 5-year survival rate was 91% and 10-year survival rate 84%.¹¹ Congestive heart failure was the commonest cause of death.

Conclusion

Although Takayasu arteritis has been recognized for more than a century, we do not seem to have been able to help our

patients that much yet. Better techniques have to be developed for managing the complications of the disease such as stenotic lesions and valvular replacement. New investigations such as positron emission tomography scans appear to be better in monitoring disease progression and therefore may lead to more accurate and targeted treatment. However, it is also important for physicians to be aware of the multiple possible forms of presentation of the disease, thereby making an early diagnosis and commencement of treatment possible, which in turn will hopefully lead to better outcomes.

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