CLINICAL VIGNETTE

Severe Aortic Regurgitation in Takayasu Arteritis

Samantha Hartnett, MD[1], Samuel Gordon, MD[2] and Sonia Shah, MD, FACC[2]

[1]Division of General Internal Medicine, Department of Medicine, Harbor-UCLA Medical Center
[2]Division of Cardiology, Department of Medicine, Harbor-UCLA Medical Center

Abstract

Severe aortic regurgitation results from aortic valve incompetence due to either primary leaflet or secondary non-leaflet pathology, resulting in symptoms of dyspnea, chest pain, and heart failure. We describe an unusual presentation of severe aortic regurgitation in a young, previously healthy woman due to Takayasu arteritis.

Case Summary

A 29-year-old female was sent to the Emergency Department from an outside cardiology office for critical findings on a transthoracic echocardiogram (TTE). She reported two weeks of progressive lower extremity edema, orthopnea, and dyspnea with minimal activity. Prior to developing symptoms, the patient felt well and had completed a strenuous 4-mile hike three weeks earlier without difficulty. She had one uncomplicated pregnancy 4 years ago. There was no history of cardiovascular disease, including hypertension.

Initial vital signs in the emergency department were significant for tachycardia with a heart rate of 120 bpm, tachypnea and hypoxia on room air requiring 3L supplemental oxygen via nasal cannula. Physical exam revealed respiratory distress, signs of volume overload, and a diastolic decrescendo murmur at the base. Discrepant blood pressures between the right and left upper extremities (160/77 vs 121/89) were noted.

Relevant labs included a mild leukocytosis, stable microcytic anemia, thrombocytosis, elevated brain natriuretic peptide, and elevated ESR/CRP. Pulmonary embolism was ruled out by CTA and the patient was admitted to Cardiology for new-onset heart failure. TTE demonstrated normal left ventricular cavity size with a reduced ejection fraction to 30-35%, severe aortic regurgitation and a mildly dilated ascending aorta. Transesophageal echocardiogram (TEE) revealed a trileaflet aortic valve with severe aortic regurgitation, primarily central, with a normal aortic root dimension and mildly dilated ascending aorta. There were no vegetations, however, thickening of the wall of the descending thoracic aorta was observed.

Given the patient's young age, elevated inflammatory markers, aortic wall thickening, and lack of other clear etiology of severe AR, the diagnosis of Takayasu arteritis was suspected. Additional imaging with a CTA of the chest revealed narrowing at the origin of the left subclavian artery and bilateral carotid stenosis.

The patient was initially treated with diuretics and right heart catheterization showed elevated left-sided filling pressures. Despite a good response to diuretic therapy, there was concern for a tenuous hemodynamic state. After a multidisciplinary discussion between Cardiology, Cardiothoracic Surgery, and Rheumatology, the decision was made to initiate pulse dose steroids and pursue urgent surgical valve replacement. Treatment with tocilizumab was considered, however, it was deemed that surgery could not be delayed. Intraoperative findings included significant aortic wall thickening and grossly normal aortic valve leaflets. Biopsy of the aortic wall confirmed the diagnosis of Takayasu arteritis.

The patient had an uncomplicated post-operative course and recovery and is followed closely by Cardiology, Cardiothoracic Surgery, and Rheumatology teams.

Discussion

This case represents not only a rare presentation of Takayasu arteritis (TA) but also an uncommon cause of severe aortic regurgitation. TA is a chronic inflammatory arteritis of the large vessels causing stenosis, dilation, and aneurysms of the aorta and it branches.[1] The disease classically progresses through three phases. The first is characterized by constitutional symptoms such as fever, malaise, arthralgia, and weight loss. Next, a vascular inflammatory phase develops with symptoms of angiodynia. Finally, the occlusive phase when the classic symptoms of TA including pulselessness develop as a result of arterial stenosis.[2] Aortic regurgitation (AR) has been reported in 25% of cases and is thought to be a later manifestation of the disease.[3] Our patient's presentation was unusual as her symptoms were entirely attributable to heart failure from AR with no preceding systemic symptoms.

Echocardiography was an important diagnostic tool in determining not only the severity of aortic regurgitation but also the etiology. Large-vessel vasculitis is the cause of AR in only 3% of cases.[4] In searching for the mechanism of AR without overt

cusp pathology, the key clue to the diagnosis of TA was the observation of wall thickening in the descending thoracic aorta on the TEE. Coupled with an elevated ESR and the patient's young age, TA was recognized as the primary underlying diagnosis. CT angiography revealed thickening of the aortic walls and stenosis of the carotid arteries and confirmed our suspicion.

Urgent aortic valve replacement in the setting of TA poses unique challenges. Considerations such as the expected response to immunosuppression and the effect of tissue friability on surgical success must be taken into account. In one case reported by de Silva, a 25-year-old female presenting with TA induced heart failure, had improvement of symptoms with just medical management.[5] Other studies have shown good outcomes with surgical management, including a study by Fields who found that long-term survival is excellent regardless of disease activity at the time of operation.[6] This is further supported by a retrospective study by Miyata who found the 15-year survival rate was increased from 43% to 67.5% following the addition of surgical intervention when more than 2 major complications (including AR) were present.[7] The presence of active inflammation may be a predictor of pseudoaneurysm formation which can occur along suture line in the setting of fragile, inflamed tissue.[8] Other potential complications following surgery include prosthetic valve detachment or progressive aortic root dilation. Therefore, pre- and post-operative steroid administration should be considered to control inflammation and limit the development of such complications.[8] Ultimately, we assessed the risk of hemodynamic collapse to outweigh the risk of surgical complications and proceeded with an aortic valve replacement.

Conclusions

Aortic regurgitation with decompensated heart failure as the primary presenting feature of Takaysu arteritis is rare. Surgical valve replacement has a good long-term outcome, though patients must be followed closely for sequelae such as prosthetic valve detachment, pseudoaneurysm formation, or progressive dilation of the aorta.

Figures

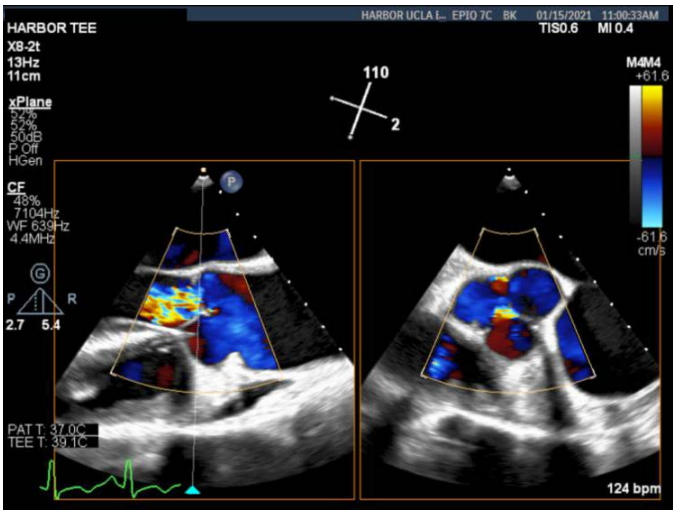

Figure 1. Transesophageal echocardiogram with color Doppler using X-plane across the aortic valve demonstrates a trileaflet valve with significant aortic regurgitation.

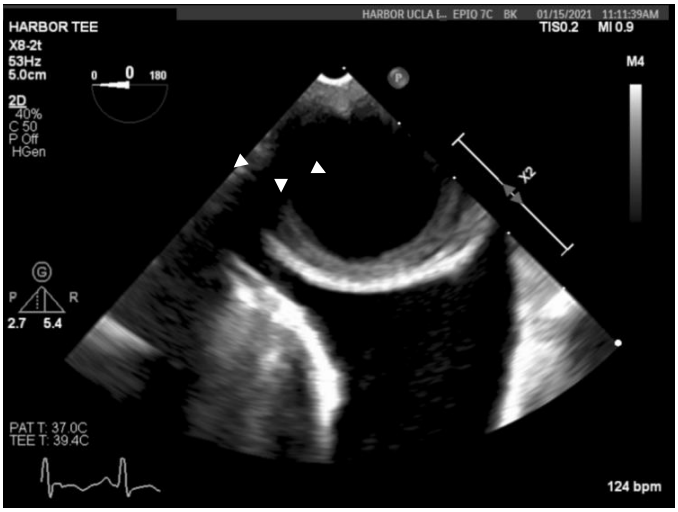

Figure 2. Transesophageal echocardiogram with 2-D imaging shows wall thickening (white arrows) around the circumference of the descending thoracic aorta.

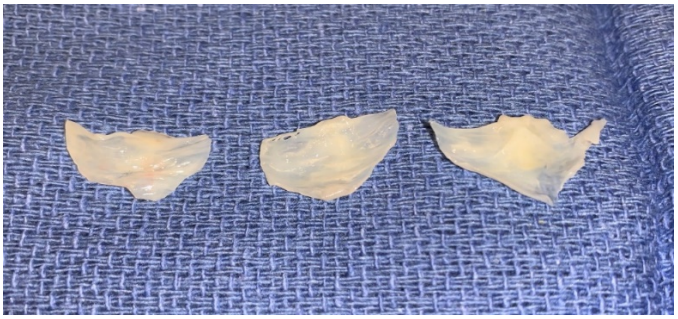
Figure 3. Gross specimen showing apparent normal aortic valve cusps after surgical removal.

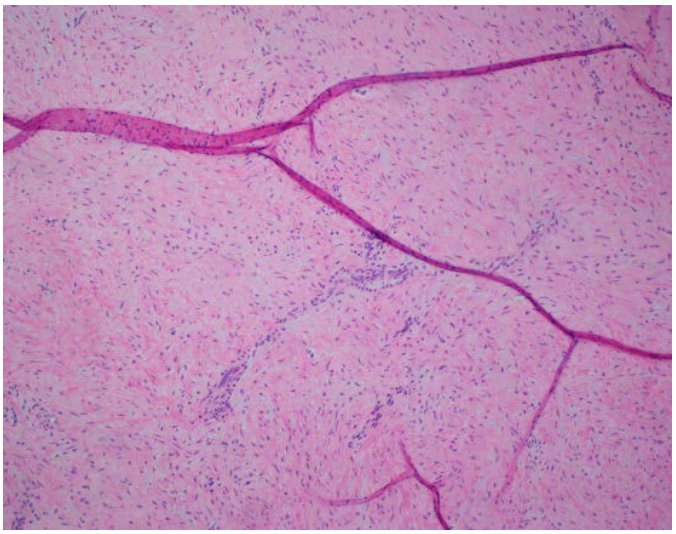

Figure 4. Pathological specimen of the aortic valve showing myxoid degeneration.

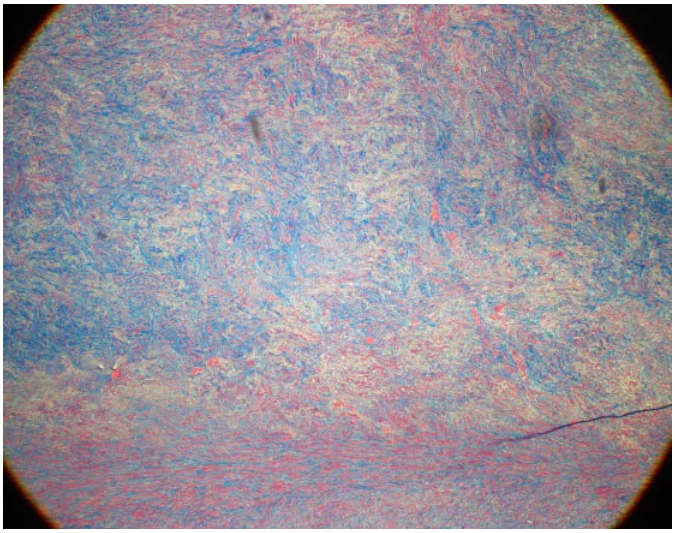

Figure 5. Pathological specimen of the aortic wall showing intimal hyperplasia, loss of smooth muscle, fragmentation of elastic tissue with replacement of fibrosis, focal lympho-histiocytic infiltrate and adventitial fibrosis suggestive of the chronic phase of Takayasu Arteritis.

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