**Complete recovery of severe left ventricular systolic dysfunction and mitral regurgitation after stenting of the descending thoracic and infra-renal aorta in a patient with Takayasu arteritis**

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**Abstract:**  
A 16-year-old girl was referred for evaluation of worsening breathlessness, limb claudication (both legs and left arm) and weight loss of three years duration. Physical examination revealed discrepancy in arterial pulses, palpable pulsations in the supra-ternal region and intercostal spaces carotid, intercostal and abdominal aortic bruit S3 gallop and pansystolic murmur of mitral regurgitation. Inflammatory markers were markedly elevated. Echocardiography showed severe left ventricular (LV) systolic dysfunction (LV ejection fraction 25) with moderately severe mitral regurgitation. On angiography, long segment tubular stenosis of descending thoracic aorta and infra-renal aorta was seen with significant narrowing of multiple branches of the aorta. Central aortic pressure was 200/78 (124) mm-Hg with peak gradients of 48 mm-Hg across the descending thoracic aorta and 40 mm-Hg across the infra-renal abdominal aorta. A diagnosis of type-V Takayasu arteritis was made using the angiographic classification. Percutaneous angioplasty and deployment of self-expanding Wallstents was performed across both stenotic segments after initial medical stabilization. Since then she has been free of lower limb claudication, and at 6 months follow-up her LV systolic function had normalized (LV ejection fraction 52) with regression of mitral regurgitation.  

**Keyword:** Takayasu arteritis, descending aorta, percutaneous transluminal angioplasty, left ventricular dysfunction.

**Introduction:**  
Takayasu arteritis is an autoimmune vasculitis that affects predominantly the aorta, its main branches and the pulmonary arteries. Congestive heart failure in Takayasu arteritis is well-known and may occur due to systemic hypertension, valvular incompetence, dilated cardiomyopathy secondary to myocarditis, coronary...
artery involvement and secondary to pulmonary hypertension [1]. There have been case reports of reversal in heart failure after balloon angioplasty in coarctation of aorta [2] or after PTA in patients with Takayasu arteritis [3].

**Case report:**
A 16-year-old girl was referred to our hospital for evaluation of worsening breathlessness (history of paroxysmal nocturnal dyspnoea and orthopnoea), limb claudication (both legs and left arm) and weight loss of three years duration. Physical examination revealed emaciation (BMI 12 kg/m²), orthopnoea, and significantly different blood pressures (mmHg) in the extremities: right arm 150/80, left arm not recordable, right leg 130/70, and left leg 130/70. Visible/palpable arterial pulsations were noted in the supra-sternal and left inter-scapular regions and over the abdominal aorta. Her heart rate and respiratory rate were 110/min and 36/min respectively. Left carotid, inter-scapular and abdominal systolic bruits with S3 gallop and pansystolic murmur in the mitral area of intensity grade 3/6 were present on auscultation. The hematological and biochemical values were within normal ranges, except for the increased erythrocyte sedimentation rate (ESR, 65 mm at 1hr) and C-reactive protein (CRP, 15.6 mg/L) and normocytic normochromic anemia (hemoglobin 9.5g/dL). The chest X-ray [Fig-1] revealed cardiomegaly (cardio-thoracic ratio-0.65) with pulmonary venous congestion. Electrocardiography [Fig-2] showed sinus tachycardia and left ventricular hypertrophy with strain pattern. Trans-thoracic echocardiography [Fig-3] showed severe global left ventricular (LV) systolic dysfunction (LV ejection fraction 25%, determined by modified Simpson method), moderately-severe mitral regurgitation (jet area/ left atrial area 37%), left atrial enlargement (dimension 4.5cm), LV dilatation (internal dimensions: diastolic 7.8cm/ systolic 6.8cm) without hypertrophy (inter-ventricular septum and LV posterior wall thickness 1.1cm each)
Fig. 3: Two-dimensional echocardiography in parasternal long-axis view showing dilated left atrium and left ventricle with mild pericardial effusion.

Anti-failure treatment (diuretics, nitrates, beta-blocker and angiotensin converting enzyme inhibitors (ACEIs) were added sequentially), and immuno-suppressive therapy (deflazacort and mycophenolate mofetil, with 1 dose of intravenous tocilizumab) was commenced after hospitalization. Respiratory distress and orthopnoea settled with standard heart failure treatment. She was taken up for high-risk aortography and percutaneous transluminal angioplasty after informed consent.

Coronary angiography, aortography and hemodynamic measurement were done. Multiple stenoses of aorta and its branches were seen. There was diffuse [Fig. 4,5] long-segment (8cm) significant (70%) narrowing of the descending thoracic aorta along with another long stenotic lesion (5cm) in the infra-renal abdominal aorta [not shown here] with stenoses of right subclavian artery (70%), left common carotid artery (50%) and celiac artery (90%). The left subclavian artery was occluded after a short stump. Coronary and pulmonary angiograms were normal. There was severe systemic hypertension - central aortic pressure was 200/78 (124) mmHg, with peak systolic pressure gradients of 48 mmHg across the descending thoracic aorta and 40 mmHg across the infra-renal abdominal aorta. A diagnosis of type V Takayasu arteritis was made using the angiographic classification [4].

Fig. 4 and 5: Digital subtraction angiography showing long-segment stenosis of descending thoracic aorta. The stenosis length was measured using a marker pig-tail catheter.

Balloon angioplasty and stenting of the diseased segments of the aorta was planned afteraortography based on the premise that relieving aortic obstruction would decrease central aortic blood pressure and increase renal blood flow and ultimately improve LV systolic function. Angioplasty and stenting [Fig. 6] was performed in the descending thoracic aorta; a self-expanding Wallstent (Boston Scientific) of size 18x90 mm was deployed. Additional angioplasty and stenting [not shown here] with another Wallstent of size 18x60 mm was performed for the stenosis of infra-renal abdominal aorta. Immediate post-procedure there was no residual gradient across the diseased segments of the aorta and central aortic pressure decreased to 187/88/120 mm Hg while femoral artery pressure increased from 112/58 (75) mmHg to 180/84 (115) mmHg.
At 6-months clinical follow up, the patient was free of lower limb claudication and orthopnoea, and her effort tolerance had improved significantly. 2D echocardiogram [Fig. 7] after 6 months showed that LV systolic function had completely normalized (LVEF 52% as measured by modified Simpson method) and there was regression of left ventricular dilatation (LV internal dimensions: diastolic 4.5cm/ systolic 3.3cm), left atrial size (dimension 3.6 cm) and severity of mitral regurgitation. However she still had persistent disease activity (ESR 88mm at 1hr, CRP 25 mg/L); aggressive immunosuppressive therapy with additional monthly doses of tocilizumab was planned.

**Fig. 6: Digital subtraction angiography of DTA after PTA with 18x90 mm Wallstent.**

**Fig. 7: Two-dimensional echocardiography in parasternal long-axis view showing regression of LA/LV dilatation at 6-months follow up.**

**Discussion:**

Takayasu arteritis can be defined as chronic inflammatory arteritis that involves the aorta, its branches and the pulmonary artery. The accepted standard for diagnosis is the American college of Rheumatology 1990 criteria [5] which have 90.5% sensitivity and 97.8% specificity. According to the new angiographic classification of Takayasu arteritis that depends on the angiographic finding and vessel involvement, the disease is classified into Type I, involving the branches of the aortic arch; Type IIa, involving the ascending aorta, aortic arch, and its branches; Type IIb, which in addition to IIa distribution involves the descending thoracic aorta as well; Type III, the descending thoracic aorta, abdominal aorta; and/or renal arteries; Type IV, abdominal aorta and/or renal arteries; and Type V, combines features of types IIb and IV [4]. This classification is useful in that it allows comparison of patients according to the vessels involved and is helpful in planning surgery, but offers little information on prognosis. Ishikawa defined clinical groups based on the natural history and complications of the disease and has prognostic significance [6]. The activity of Takayasu's arteritis can be measured by the presence of the symptoms, typical angiographic findings, and documenting the disease activity [elevated ESR, CRP, interleukin -6, regulated on activation, normal T cell expressed and secreted (RANTES) etc] [7, 8]. Disease activity can also be measured by CT, MR, and (18F) FDG-PET imaging in a non-invasive manner. The intensity of MR enhancement of the arterial wall and [18F] FDG-PET show a positive correlation with both the ESR and CRP levels, therefore, they may be correlated with the disease activity and so represent useful diagnostic indicators [9,10 ,12]. The standard immunosuppressive treatment for Takayasu arteritis includes steroids (such as deflazacort) combined with methotrexate, azathioprine, cyclophosphamide and mycophenolate mofetil [11]. Recent reports have indicated that the interleukin-6 receptor antagonist monoclonal antibody tocilizumab can be tried when the patients experience a relapse of disease activity in spite of steroid treatment. Anti-hypertensive therapy is necessary if systemic hypertension is present. The operative treatment consists of open surgery or percutaneous intervention.
Surgery comprises of reconstruction of aorta with resection of stenotic segment, interposition of graft or aorto-aortic bypass. Besides its associated morbidity, surgical bypass is often complicated by graft occlusion, anastomotic site aneurysm [13]. Angioplasty offers a less invasive, cost-effective, and safe method for relief of stenotic lesions in patients with Takayasu’s arteritis [14]. Our patient had severe left ventricular systolic dysfunction, moderately severe mitral regurgitation, elevated central aortic pressure and highly active disease (markedly elevated ESR and CRP at admission). Takayasu arteritis can lead to a dilated cardiomyopathy like picture due to afterload mismatch caused by severe elevation of central aortic blood pressure or due myocarditis. Other causes of LV dysfunction in Takayasu arteritis are coronary artery involvement and aortic regurgitation, but these were absent in this patient. Presence of myocarditis cannot be ruled out in this patient because endomyocardial biopsy was not performed; the mild posterior pericardial effusion seen on initial transthoracic echocardiography could be due to myopericarditis or due to heart failure itself. The fact that there was significant clinical improvement even though the disease remained active (as per inflammatory markers, despite immunosuppressive medication) may indicate that LV dysfunction was more related to the central aortic hypertension (that was relieved by percutaneous intervention) than to myocarditis. However, a high index of suspicion of myocarditis is necessary in such patients because stopping the ACE inhibitor or betablocker or premature tapering of immunosuppressive medications could prevent recovery or precipitate relapse of LV systolic dysfunction; hence it is imperative that all these medications are continued well beyond the time when LV function normalizes and disease remission is achieved.

**Conclusion:**

In this young patient with symptomatic type-V Takayasu arteritis and significant central hypertension and LV systolic dysfunction, standard immunosuppressive therapy along with angioplasty and stenting of the descending thoracic aorta and infra-renal abdominal aorta lead to complete recovery of left ventricular systolic function with regression of left ventricular dilatation and mitral regurgitation.

**Bibliography**


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