



## Case Report

### Type IV Takayasu's Arteritis: An Unusual Case Presentation in a Young Indian Woman

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#### ABSTRACT

Takayasu's arteritis (TA) is a rare disorder involving chronic inflammatory arteritis affecting aorta and its main branches, which may affect multiple organs. Herein, we present a case of a young woman referred with hypertension and bilateral renal artery stenosis for further evaluation, and was diagnosed as a case of Type IV Takayasu's arteritis.

**KEYWORDS:** Takayasu's arteritis, Indian women.

#### INTRODUCTION

Takayasu's arteritis (TA) is a rare disorder involving chronic inflammatory arteritis affecting aorta and its main branches.[1] TA is more common in South-East Asian regions with a worldwide distribution, and is the most common cause of renovascular hypertension.[2] TA presents in younger adults commonly in the 2<sup>nd</sup> or 3<sup>rd</sup> decade of life with symptoms such as fever, arthralgias, weight loss, arm claudication and hypertension. Laboratory investigations reveal anemia and increase in erythrocyte sedimentation rate (ESR), and angiograms reveal stenosis at various levels of aorta and its branches.[3]

As per American College of Rheumatology criteria, TA is diagnosed in presence of any 3 of the following 6 criteria - onset at age less than or equal to 40 years, claudication of an extremity, decreased brachial artery pulse, greater than 10 mm Hg difference in systolic blood pressure between arms, a bruit over the subclavian arteries or the aorta, and arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities.[4] TA is associated with significant comorbidities such as hypertension, congestive heart failure, claudication, visual symptoms, cerebrovascular accidents, etc. The mortality rate has been estimated to be about 15%, with a 5-year survival rate of 84%, with main cause of death being attributed to

congestive heart failure.[5] The present report is a case of young woman referred to our centre who was evaluated and diagnosed as type IV TA.

#### CASE REPORT

An 18 year old Indian woman with a diagnosis of bilateral renal artery stenosis on vascular Doppler was referred to our centre for renal angiogram. On eliciting history of the patient, she had fever since 1 month, tightness in chest and palpitations since 15 days and blurring of vision since 3 days. She was suffering from hypothyroidism since 5 years and hypertension since 1 year. There was no history of tuberculosis, inflammatory bowel disease, complaints of limb ischemia and similar disease in family. On general examination, her pulse rate was 100 beats per minute, blood pressure measured on the left upper limb was 190/100 mm Hg, on the right upper limb was 160/90 mm Hg and on both lower limbs it was 190/100 mm Hg. Rest of general and systemic examination was found to be within normal limits. The patient was further investigated for fever and hypertension.

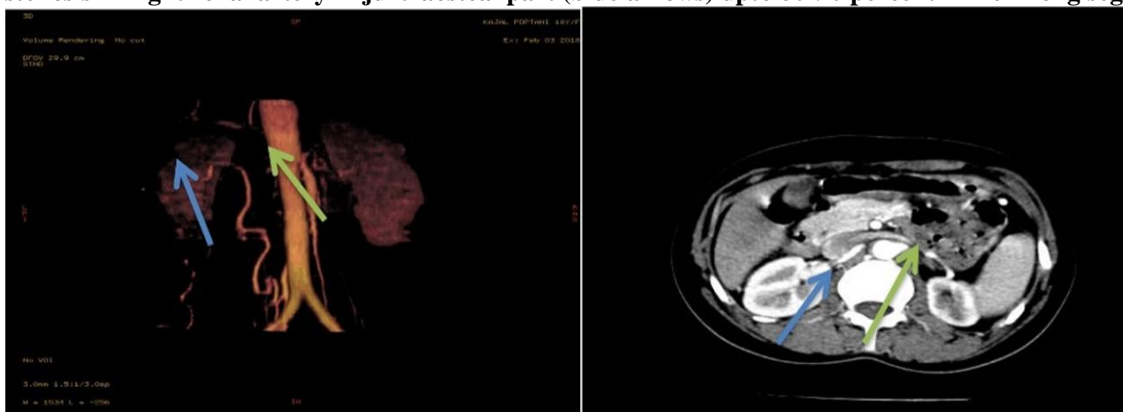
Fundal examination revealed lesions which suggested a differential diagnosis of grade IV hypertensive retinopathy or resolving vasculitis. Complete blood count revealed a total leucocyte count of 2100/mm<sup>3</sup>, the liver and renal function tests were within normal limits, Widal test was positive. Urine routine examination was normal. Erythrocyte

sedimentation rate (ESR) at the end of 1 hour was 42 mm and C-reactive protein (CRP) was 1.2 mg/dl. Anti-neutrophil antibodies (ANA) were in normal range. A 2D ECHO was suggestive of concentric left ventricular hypertrophy with an ejection fraction of 60%. Coarctation of aorta was ruled out. Patient underwent contrast enhanced computed tomography (CECT) of the abdomen and thorax. The aortogram (Figures 1-3) was suggestive of bilateral renal artery stenosis along with stenosis of superior mesenteric artery. The thoracic and abdominal aorta along with major branches were normal. These features were suggestive of Type IV Takayasuarteritis.

As ESR and CRP were raised, right basal sub-segmental consolidation was noted on CT, the stenting of renal artery

was deferred. She was treated with ceftriaxone 1 gm intravenously twice a day for 14 days and tablet azithromycin 500 mg once daily orally for 7 days. She was started on tablet prednisone 1 mg/kg for 6 weeks. For management of hypertension, she was started on a fixed dose combination of amlodipine 5 mg and atenolol 50 mg twice a day, tablet tosaemide 10 mg plus spironolactone 50 once in morning and tablet prazosin 5 mg at night. At 6 weeks of treatment, she was afebrile and the blood pressure reduced to 160/90 mm Hg on left upper limb, and 120/70 mm Hg on right upper limb. However, the ESR was found to be raised (40 mm at end of 1 hour). Hence, it was decided to continue the patients on steroids, antihypertensives and stenting of renal artery was further deferred.

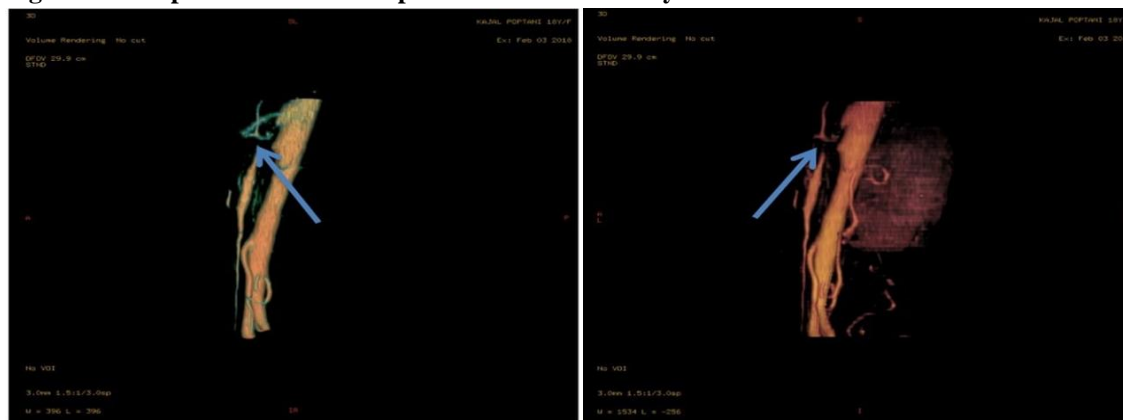
**Figure 1: Severe non-segmental stenosis in left renal artery (green arrows) upto 70-80 percent with severe short segment stenosis in right renal artery in juxtaaosteal part (blue arrows) upto 80-90 percent in 1 cm long segment.**



**Figure 2: Near complete occlusion of coeliac trunk**



**Figure 3: Complete occlusion of superior mesenteric artery**



**Figure 4: Uninvolved arch and its branches and thoracic**



## DISCUSSION

TA is a rare disease with varying incidences globally of 0.4-2 per million people in different hospital and registry based studies.[6] Published literature on TA has demonstrated higher females: males ratio ranging from 1.58 in India[2] to 4.33 - 29 in North America[3,7]. TA is a multi-systemic disorder posing a challenge in diagnosis especially in the initial stages, owing to lack of serologic test and presentation with general constitutional symptoms.[8] The patient in this case was suspected for TA as the patient was a young female, with high blood pressure showing differences on systolic blood pressure on upper limbs of 30 mm Hg, and renal artery stenosis.

On evaluation for aorta using CECT, the branches of abdominal aorta i.e. superior mesenteric artery and bilateral renal arteries, showed stenosis which satisfied 3 of the 6 criteria for diagnosing TA. The patient was diagnosed as a rare case of type IV TA. TA is also known as 'pulseless disease', but in this case the pulse of the patient could be felt and was on upper limit, and thus, was an atypical presentation of TA. Diminished or absent pulses are noted in 84-96% patients with limb claudication, vascular bruits in 80-94%[1], which were not observed in the present case. Fever, as seen in current case, as a presentation of TA is very rarely reported.[9,10]

The angiographic findings of TA have been classified in 6 types - Type I branches from aortic arch, Type IIa ascending aorta, aortic arch, and its branches, Type IIb ascending aorta, aortic arches and its branches, thoracic descending aorta, Type III thoracic descending aorta, abdominal aorta and/or renal arteries, Type IV abdominal aorta and/or renal arteries, Type V showing combined features of types IIb and IV.[11] The patient described in the case involved only the branches of the aorta and not the abdominal aorta, which is distinct from the traditionally described cases of TA with involvement of abdominal aorta in addition to its branches.

In Indians, type IV is more common compared to Japanese population who have a predominance of type I and IIa.[11] Renal artery stenosis in TA is sequential to acute inflammatory arteritis and lymphocytic infiltration, thickening of intima, destruction of elastic tissue, fibrosis and patchy luminal narrowing, eventually leads to blood flow reduction and reno-vascular hypertension.[12] However, the etiology and pathogenesis of TA remains

unanswered. Immunosuppression forms the mainstay treatment approach for TA. In active disease, treatment of high dose corticosteroid such as prednisolone (1 mg/kg/day) leads to remission, however, relapses can occur.

Other conventional immunosuppressants including methotrexate, azathioprine, mycophenolatemofetil, cyclophosphamide, and leflunomide have shown promising results. Those still resistant or intolerant are treated with biologicals like TNF inhibitors, rituximab and tocilizumab. In life-threatening conditions due to short-segment arterial stenosis, revascularisation using stenting or balloon angioplasty and in long-segment arterial stenosis, bypass grafting are performed.[13]

Prognosis of TA depends on presence of major complications including aortic regurgitation, retinopathy and aneurysm formation, rate of disease progression, and high ESR.[12] The patient in the present case had retinopathy and high ESR, and is a complicated type TA. The 15-year survival for complicated and uncomplicated TA have been reported to be 66.3% and 96.3%, respectively.[14] Follow-up of the patient for a longer duration will, thus, be required in the current case.

## CONCLUSION

A young female presenting with hypertension and renal artery stenosis should be thoroughly investigated to rule out TA as a differential diagnosis. As observed from the patient in this study, the presentation of TA can be complex and atypical. It is important to categorize the patient for the type of TA and presence of complications. Technical advances assist in better localizing the affected parts of the aorta in a TA patient.

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