A Rare Variety of Takayasu Arteritis
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Abstract
We diagnosed a case of Takayasu arteritis (TA) involving subclavian arteries, the aorta, superior mesenteric artery and renal arteries presenting with stenotic, occlusive, and aneurysmal lesions along with mural thrombus, which responded well to ATD and steroids. We report this case as a rare combination of vascular lesions in a patient with a relatively rare variant of TA.

Introduction
TA is a chronic granulomatous panarteritis of large sized arteries, classically involving the aortic arch, but one third of the cases also affect the remainder of the aorta, its branches, and pulmonary arteries. We report a case of TA type V with extensive and varied involvement of the aorta and its branches that responded well to antituberculosis drugs and steroids.

Case Report
A 28 year old unmarried non-diabetic, hypertensive lady, presented with the chief complaints of pain and numbness of both left limbs for four months and weakness of the same limbs along with low grade fever for the last one month. She was on Amlodipine 5 mg daily past 2 months and had been exposed to TB in her family 8 years back.

The patient was cachectic (BMI 17.3 kg/m²) and febrile on admission. The pulse rate was 82/min and radial and brachial pulsations were well palpable on the right side alone. Similarly, femoral, popliteal and dorsalis pedis arteries were palpable on the right side but not on the left. However, both carotid arteries were well palpable. The BP was 160/90 mmHg over the right brachial artery and 176/100 mmHg over the right popliteal artery. A longitudinal tubular mass with expansile pulsations involving the umbilical and hypogastric areas was appreciated with audible bruit over it. The power of left upper and lower limb was 4/5 in both proximal and distal group of muscles with normal tone without sensory involvement and preserved deep tendon reflexes.

The clinical examination was otherwise unremarkable. A provisional diagnosis of large vessel vasculitis with abdominal aortic aneurysm was entertained and investigations sent accordingly.

CBC revealed normal WBC counts, Hb 8.8 gm/dL, a microcytic hypochromic picture and an ESR of 120 mm at the end of one hour. Chest X-ray suggested superior mediastinal widening. Abdominal USG showed abdominal aortic aneurysm involving the entire length of the abdominal aorta, extending through the left common iliac artery into the left external iliac artery. The maximum diameter of the aneurysm was 6cm and effective luminal diameter was 2cm. Kidney sizes were normal. The diameter of induration on Mantoux test was 40mm. Sputum was unavailable and the Echo-Doppler study documented LVH and grade I diastolic dysfunction. ANA was negative by the Hep2 method.

CT angiography of aorta and its branches showed diffuse mural thickening with wall irregularity in the left subclavian artery and left axillary artery, chronic total occlusion of 2nd and 3rd parts of the left subclavian artery and tight stenosis of the left axillary artery (Figure 1). Marked tortuosity

Fig. 1: 3D CT angiography showing normal ascending and arch of aorta proximal to origin of left subclavian artery, long segment total occlusion of 2nd and 3rd parts of the left subclavian artery, tight stenosis of left axillary artery and focal moderate stenosis near origin of the coeliac trunk and total occlusion at origin of superior mesenteric artery

Fig. 2: CT angiography showing marked tortuosity and fusiform aneurysmal dilatations involving the entire descending thoracic and abdominal aorta, with acute angulations just above the aortic hiatus and at the infrarenal aorta

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Table 1: Common clinical manifestations of TA

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<tr>
<th>Features</th>
<th>Percentage</th>
<th>Features</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Constitutional symptoms</td>
<td>66%</td>
<td>Hypertension</td>
<td>43%</td>
</tr>
<tr>
<td>Diminished pulses</td>
<td>88%</td>
<td>Aortic regurgitation</td>
<td>33%</td>
</tr>
<tr>
<td>Bruits</td>
<td>77%</td>
<td>Renal artery stenosis</td>
<td>26%</td>
</tr>
<tr>
<td>Pain in extremities</td>
<td>69%</td>
<td>Cerebrovascular accident</td>
<td>18%</td>
</tr>
<tr>
<td>Claudication</td>
<td>48%</td>
<td>Pulmonary hypertension</td>
<td>12%</td>
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Takayasu arteritis with extensive involvement of the aorta and its branches, with mural thrombus and aortic dissection, related to a Tuberculous etiology. Opinion from the Department of Cardio-thoracic and Vascular Surgery ruled out any operative intervention in such extensive vascular involvement. The patient was put on Category I anti-tubercular therapy along with oral prednisolone 40 mg daily for the initial 4 weeks, followed by tapering off over the next month. Fever subsided rapidly and the patient was put on physiotherapy and discharged after 3 weeks of hospitalisation. Six months later, there was substantial improvement of her left sided weakness with an appreciable increase in her claudication distance but no return of left sided pulses. The ESR at this time was 19 mm at the end of 1 hour.

Discussion

TA was first reported from Japan in 1908 and is prevalent in Asian populations. TA is a disease of young women (Male : Female 1 : 8, age of onset 25-30 years) and is the commonest cause of renovascular hypertension in India.1 The aortic arch is more involved in Japan while the involvement of the abdominal aorta is more in Indian and Korean patients.2 Common clinical and imaging features of TA are enlisted (Table 1).3

Our patient fulfilled the American College of Rheumatology criteria of TA (Table 2).4 The NIH has defined active disease as new onset or worsening of at least two of the following four features: (i) signs and symptoms of vascular inflammation or ischemia (claudication, decreased or absent pulses or blood pressure in the extremities, bruits or carotidynia); (ii) elevated ESR; (iii) angiographic abnormalities; and (iv) systemic symptoms like fever, polyarthralgia and polymyalgias not attributable to another disease. Vascular lesions in TA may be stenotic (93%), occluded (57%), dilated (16%), or aneurysmal (7%).5 This patient had stenosis, occlusion, and aneurysm, in addition to intra mural thrombus, a combination rarely seen in the literature. The Ishikawa clinical classification of Takayasu arteritis describes 4 groups depending on the number and severity of complications, and our patient could be categorized into Ishikawa Group 4.6 According to the new angiographic classification of TA as adopted at the Takayasu conference 1994, the aortic arch and its branches are mainly involved in Japanese patients (type I, IIa). But there was controversy regarding commonest type in India. According to few authors it is type IV7 and according to few it is type V.8 But a recent retrospective study clearly showed type V is the commonest type in India.9 Type III is the most common type found in south-east Asia and Africa and is called as ‘middle aortic syndrome’. Our patient had type V TA which is common in India.

The causal association of TA with TB has been much discussed. A 65 kDa heat shock protein (HSP) is a major immunogenic component of M. tuberculosis and expression of HSP has been shown to be strongly induced in the aortic tissue through molecular mimicry. It is possible that TA is caused by antibodies, generated following exposure of human host to bacterial HSP may cross-react with the human homologue of HSP 65, which is expressed on the surface of stressed endothelial cells. This interaction with the endothelial HSP may initiate an immune response responsible for the subsequent lesion.10,11

Higher frequency of positive tuberculin tests in TA patients than in general population were described in literature.12 Recently it was shown that skin delayed hypersensitivity to PPD with induration over 10 mm may be as frequent in TA as in patients with extrapulmonary tuberculosis (92.5% & 89% respectively).13

There is no correlation between the size of induration and likelihood of current active TB disease. But induration size of more than 15 mm are unlikely.
due to previous BCG vaccination. Latent TB infection is considered for any BCG vaccinated person when skin test is 10 mm or greater and was in contact with TB infected person or born or lived in a high TB prevalent country or continually exposed to population where TB prevalence is high. With this background Anti TB Drug administration was justified and further supported by symptomatic improvement of this patient.14

Anti-TB drugs (ATD) along with steroids have been used by authors across the Indian subcontinent in treating TA with fever and constitutional symptoms, especially if exposure to TB can be documented.15 The European League Against Rheumatism (EULAR) recommends initial high doses of glucocorticoids for disease control and an immnosuppressive agent (methotrexate, cyclophosphamide, azathioprine or mycophenolate mofetil) as an adjunctive therapy.16

### References


### Table 3: New angiographic classification of TA (Takayasu conference, 1994)16

<table>
<thead>
<tr>
<th>Type</th>
<th>Extent of vascular involvement</th>
</tr>
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<tbody>
<tr>
<td>I</td>
<td>Branches of the aortic arch</td>
</tr>
<tr>
<td>IIA</td>
<td>Ascending aorta, aortic arch, and branches of the aortic arch</td>
</tr>
<tr>
<td>IIB</td>
<td>Ascending aorta, aortic arch, and its branches and thoracic descending aorta</td>
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<tr>
<td>III</td>
<td>Thoracic descending aorta, abdominal aorta, and/or renal arteries</td>
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<tr>
<td>IV</td>
<td>Abdominal aorta and/or renal arteries</td>
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<tr>
<td>V</td>
<td>Features of types IIB and IV</td>
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### Conclusion

The purpose of documenting this case was to highlight the extensive involvement of the aorta (Type V), the variety of vascular changes that we found in a single patient (Ishikawa Group 4) and the strong causal association our patient had with a tubercular etiology. More importantly, this case possibly redefines the role of ATD in all patients diagnosed with TA in the Indian subcontinent.