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Unmasking takayasu arteritis-lessons to reckon

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Abstract
Takayasu arteritis is a rare type of large-vessel arteritis that primarily affects the aorta and its major branches. Cases of TA are under reported and under diagnosed in India. This case highlights the enigmatic nature of TA, with TIA as the presentation, while on treatment and reiterates that a high index of suspicion is required in clinical practice to make an early diagnosis of the disease. Takayasu arteritis patients constitute a management challenge because treatment options vary, depending on the stage of the disease at the time of its diagnosis. The pickup rate of TA is high in patients with suspected diagnosis. Here we present a case of a 24 year old female patient where although the clinical presentation was blurring of vision in the first attack and motor aphasia the second time suggestive of TIA, a high index of suspicion lead to the diagnosis of Takayasu's and appropriate treatment initiated at the right time lead to recovery.

Keywords: Takayasu arteritis, blurring of vision, TIA (Transient ischemic attack)

Introduction
Takayasu arteritis is a rare type of large-vessel arteritis that primarily affects the aorta and its major branches. Cases of TA are under reported and under diagnosed in India.
Worldwide incidence is estimated at 2.6 cases/million/year with the majority of cases being reported from south-east Asia and India [1].
In adults approximately 80% of patients with TA are women [2, 3]. This case highlights the enigmatic nature of TA, with TIA as the presentation, while on treatment and reiterates that a high index of suspicion is required in clinical practice to make an early diagnosis of the disease.

Case Report
A 24 year old female, with no history of abortions was referred to our hospital with complaints of blurring of vision of both eyes with episodes of complete loss of vision lasting few minutes for last 8 months, accompanied by recurrent falls secondary to imbalance and buckling of knees along with dysphagia and dysarthria since 6 months. She had also history of weakness with pain on exertion in both upper limbs. On examination peripheral pulses were weakly palpable in lower limbs but not palpable in upper limbs with unrecordable blood pressure and popliteal blood pressure of 190/100 mm hg. Both carotids were weakly palpable with audible bruit, with bilateral ocular ischemic syndrome.
Systemic evaluation showed generalized spasticity, exaggerated reflexes with extensor plantar, a right renal bruit, otherwise normal. Investigation wise, patient had a normal hemogram with elevated acute phase reactants-CRP-15mg/l and ESR-60 mm at the end of 1 hour. CT angiogram revealed a saccular aneurysm of aorta and pulmonary artery with thrombosis of left subclavian artery and short segment of right common carotid artery [Figure 1 and 2]. Renal Doppler showed complete right renal artery Stenosis. ANA and dsDNA was negative. Diagnosis of Takayasu was made based on the Ishikawa Criteria as the patient fulfilled the obligatory criteria and 1 major with 2 minor criteria [Table 1] and ACR (American College of Rheumatology) criteria with fulfillment of 3 criteria [Table 2]. She was started on steroids, antihypertensives and antiplatelets. She recovered and was discharged. 2 months later, on steroids, she presented with acute onset of motor aphasia with a BP of 200/100 and normal systemic evaluation. Her blood investigations were normal and a TIA was diagnosed as she recovered within 24 hours. Her antihypertensives were tapered up and advised nephrectomy/stenting after the flare up resolved. She was started on methotrexate and she recovered, was discharged and is on regular follow up.

Discussion
Takayasu's disease is seen in a wide geographic area, mainly in Asia and Africa. It is an autoimmune disease involving the arterial walls of large arteries, causing panarteritis [1].
The clinicalcourse of the disease is divided into an early active inflammatory phase and late chronic phase. The active phase lasts for weeks to months and may have a remitting and relapsing course. It is characterized by systemic disease with symptoms of fever, general malaise, night sweats, loss of appetite, weight loss, headaches, dizziness, arthralgia, skin rashes [3].
Evidence of vessel inflammation such as tenderness along arteries, bruits, and aneurysm may point to the diagnosis of TA [Table 3]. The late chronic phase is the result of arterial stenosis and/or occlusion and ischemia of organs. Its clinical manifestations are varied and related to the location of arterial lesions [4].
In 13.5% to 33% TA patient may present with visual loss [5].
Suspected TA mandates vascular imaging. The intraarterial angiography has been largely replaced by computed tomography angiography or magneticresonance angiography (MRA). Treatment of TA is based on the use of immunosuppressant such as prednisone and/ormethotrexate to decrease or eliminate inflammatory activity. Anti-inflammatory therapy can lead to a dramatic improvement in TA. The 5-year survival rate in adults is as high as 94% [6].
In the presence of symptomatic stenotic or occlusive lesions,

endovascular revascularization procedures like bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty, or stent placement should be takeninto consideration [7]. Both, surgical and endovascular, treatments become risky and achieve poorer outcomes, if they are undertaken during a period of inflammatory activity [8].

Fig 1: Ishikawa Criteria

Obligatory criterion
Age at disease onset <40 years or at onset of characteristic signs and symptoms of 1 month duration
Major criteria
Involvement of left mid-subclavian artery
Involvement of right mid-subclavian artery
Minor criteria
Raised ESR
Carotid artery tenderness
Hypertension
Aortic regurgitation or annuloaortic ectasia
Pulmonary artery lesions
Left mid-common carotid lesion
Distal brachiocephalic trunk lesion
Descending thoracic aorta lesion
Abdominal aorta lesion
Presence of two major criteria or one major and two or more minor criteria or at least four of the nine minor criteria (representing vessel involvement) is consistent with a diagnosis of Takayasu arteritis, with a sensitivity of 84% and specificity of 95%

ESR: Erythrocyte sedimentation rate

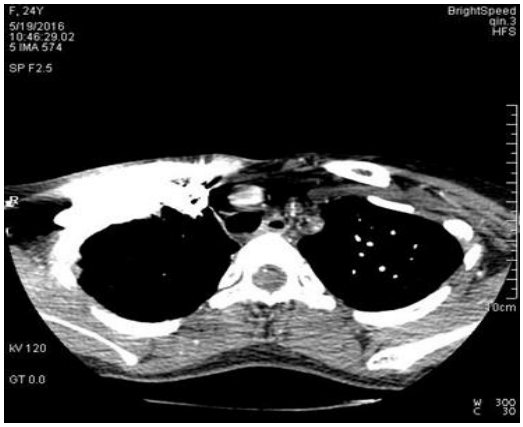
Fig 2: ACR Critertia

1. Age at disease onset <40 years: Development of symptoms or findings related to Takayasu arteritis <40 years of age
2. Claudication of extremities: Development and worsening of fatigue and discomfort in muscles of ≥1 extremity while in use, especially the upper extremities
3. Decreased brachial artery pulse: Decreased pulsation of one or both brachial arteries
4. Blood pressure difference >10 mm Hg: Difference of >10 mm Hg in systolic blood pressure between arms
5. Bruit over subclavian arteries or aorta: Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta
6. Arteriogram abnormality: Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental

The presence of >3 criteria is consistent with a diagnosis of Takayasu arteritis, with a sensitivity of 91% and specificity of 98%

Fig 3: Vessel Involvement and Symptoms

The vessels involved	Clinical features
1. Aortic branches	Malaise, decreased or absent pulse of upper extremities, dysfunction of upper extremities, headaches, dizziness, vision and orientation disturbances, syncope[12,13]
2. Aortic arch	Congestive heart failure, aortic valve insufficiency, arterial hypertension[14,15,16]
3. Coronary arteries	Ischemic heart disease, myocardial infarction[17,18]
4. Pulmonary arteries	Chest pain, dyspnea, coughing, hemoptysis, congestive heart failure[19]
5. Abdominal aorta or celiac trunk	Ischemia of the stomach and intestines, abdominal pain, nausea, vomiting[20]
6. Renal arteries	Arterial hypertension, chronic renal failure[21,22,23]



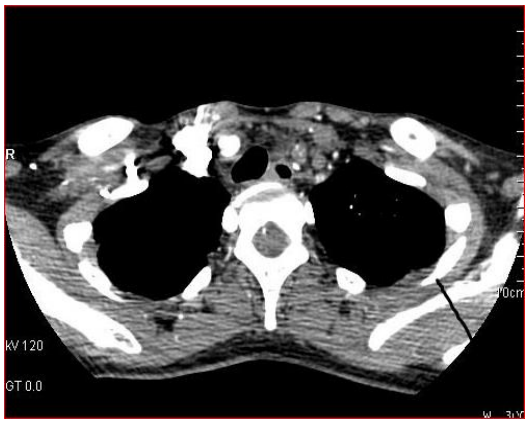


Fig 1&2: CT angiogram revealed a saccular aneurysm of Aorta and pulmonary artery with thrombosis of left subclavian artery and short segment of right common carotid artery

Conclusion

Takayasu arteritis patients constitute a management challenge because treatment options vary, depending on the stage of the disease at the time of its diagnosis.

The pickup rate of TA is high in patients with suspected diagnosis. The current case illustrates such an example. Though clinical presentation was blurring of vision in the first attack and motor aphasia the second time suggestive of TIA, a high index of suspicion lead to the diagnosis and appropriate treatment.

Renovascular hypertension should be appropriately managed with antihypertensives and stenting as required in a case of TA. Neurological manifestations are a rare presentation in TA but must be kept in mind in a young female. Atypical presentations should be kept in mind while diagnosing TA.

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