Case Report

Takayasu Arteritis

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ABSTRACT

This report describe a 26 year old female who presented with gradual dimness of vision, syncopal attacks, and arm claudication. She was diagnosed as a case of Takayasu arteritis. Takayasu arteritis or "pulseless" disease is a rare idiopathic, chronic granulomatous vasculitis that affects aorta and its major branches.

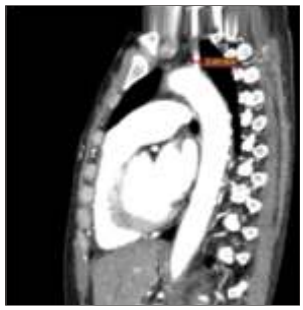
Introduction :

Takayasu's arteritis (TA), also known as aortoarteritis and pulseless disease, is a rare condition. It is a form of granulomatous arteritis, which affects large- and medium-sized arteries, primarily the aorta and its large branches as well as proximal portions of pulmonary, coronary, and renal arteries. Initially, there are mononuclear cell infiltrations in the adventitia and granulomas with Langerhans cells in the media, followed by disruption of the elastin layer and subsequent massive medial and intimal fibrosis. These lesions result in segmental stenosis, occlusion, dilatation, and aneurysmal formation in the affected vessels. Diminished or absent pulses are present in 84-96% of patients, associated with limb claudication and blood pressure discrepancies between the two arms. The symptoms are secondary to the involved artery, and it may evolve into a life-threatening condition. Involvement of the carotid artery results in ophthalmic artery hypoperfusion and causes ocular ischemic syndrome.

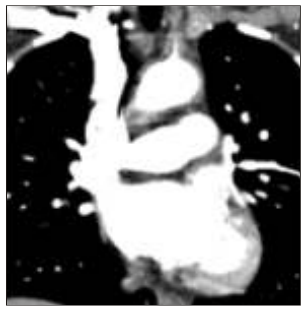
Case Report :

26 year old female had a gradual decline in visual acuity over the past 3 months in both eyes. She had past history of generalised fatigue, pain and tingling sensation in both upper and lower limb resembling claudication for the last 3 years.

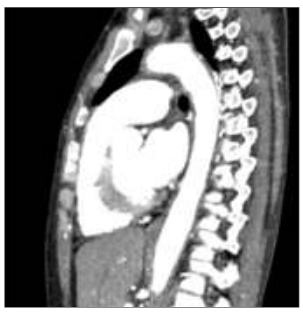
CT AORTOGRAM



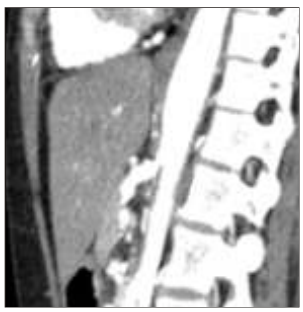
Narrowed Brachiocephalic Trunk with Wall Irregularity



Narrowing of Left Common Carotid



Thickening involving Proximal Abdominal Aorta



Severe Ostial Narrowing of Celiac Artery

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On examination, Brachial and radial arterial pulses of left side were absent on palpation. Right sided radial artery was absent and brachial artery was palpable but was very feeble. Her Blood Pressure was 150/90 in both lower limbs and 70 systolic in right arm but non recordable on left arm. There was bruit over right internal carotid artery. Visual acuity was 6/36 in the right eye and counting fingers at 2 meter in the left eye.

Her ESR was 95 mm at the end of one hour and serum C-reactive protein level of 5.4 mg/dl. LFT, KFT, Thyroid Profile were Normal. CT Aortogram shows circumferential wall thickening in aortic arch, distal thoracic aorta and proximal abdominal aorta.Brachiocephalic trunk narrowed in calibre and showed wall irregularity; Right common carotid and right subclavian arteries appear narrowed in caliber with circumferential wall thickening. There was narrowing of left common carotid artery for a length of 3.7 cm from its origin. Similarly left subclavian artery was narrowed for a approximate length of 2.5 cm. from its origin. Left vertebral artery shows focal narrowing at its origin. There was severe focal ostial narrowing (2.8 mm) noted at origin of celiac artery. SMA appears severely narrowed. On the basis of clinical manifestations and angiographic findings, the diagnosis of Takayasu Aortoarteritis was made.

The patient was given three doses of methylprednisolone and followed by oral steroids on a tapering basis. We consulted with vascular surgeons who advised conservative management as there was multiple vessel involvement.

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. The American Rheumatological Society considers three of the following six criteria necessary for a definite diagnosis of Takayasu's disease :

1. Onset before 40 years
2. Claudication of the extremities
3. Decrease in the brachial pulse in one or both arms
4. Difference of 10 mm Hg or more in blood pressure measured in both arms.
5. Audible bruit on auscultation of the aorta or subclavian artery
6. Narrowing at the aorta or its primary branches on arteriogram.

The current patient met all six criteria. Our patient was a 26 years old female. Though TA is predominantly a disease of young adults in the second and third decades of life, but not uncommon in childhood and in adults older than 40 years[5,6,7]. The youngest patient described was 6 months old and the oldest one was 75 years. Although females are more likely to be affected by this TA but males are also affected. In adults approximately 80% of patients with TA are women[6]. The female-to-male ratio varied from 9:1 in reports from Japan, 6.9:1 in Mexico, to 1.2:1 in Israel.

Clinical manifestations of TA are nonspecific. The clinical course 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 characterised by systemic disease with symptoms of fever, general malaise, night sweats, loss of appetite, weight loss, headaches, dizziness, arthralgia, skin rashes, etc. The acute phase does not occur in all patients, but constitutional symptoms are often seen in children with TA. It should be highlighted that the correct diagnosis of TA is seldom made in the early phase. Evidence of vessel inflammation such as tenderness along arteries, bruits, and aneurysm may point to the diagnosis of TA[5]. The late chronic phase is the result of arterial stenosis and / or occlusion and ischemia of organs

Type	Vessel involvement
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Type I	Branches from the aortic arch
Type IIa	Ascending aorta, aortic arch and its branches
Type IIb	Ascending aorta, aortic arch 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	Combined features of types IIb and IV

Our patient presented with visual loss, malaise, absent pulses of left upper extremities, dizziness and syncope as she had occlusion and stenosis of branches of aorta. In 13.5% to 33% TA patient may present with visual loss.

Suspected TA mandates vascular imaging. While the intra-arterial angiography still remains the standard for diagnosis and evaluation of Takayasu arteritis, it has been largely replaced by computed tomography angiography or magnetic resonance angiography (MRA). Treatment of TA is based on the use of immunosuppressant such as prednisone and / or methotrexate 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%.

In the presence of symptomatic stenotic or occlusive lesions, endovascular revascularization procedures like bypass grafts, endarterectomy, percutaneous transluminal angioplasty, and stent placement should be taken into consideration[3]. The status of such treatment is controversial in the literature. Despite providing short-term benefit, endovascular revascularization procedures are associated with a high failure rate in patients with Takayasu's arteritis. Published results suggest that these procedures should be undertaken with great care and be reserved for specific indications. Both, surgical and endovascular, treatments become risky and achieve poorer outcomes, if they are undertaken during a period of inflammatory activity.