

CASE REPORT OF TAKAYASU'S ARTERITISAvinash Gupta¹, Divya Bagoria², Jaya Pamnani³, Sumitra Choudhary⁴¹Associate Professor, Department of Radio-diagnosis, JLN Medical College, Ajmer.²2nd Year Resident Doctor, Department of Radio-diagnosis, JLN Medical College, Ajmer.³2nd Year Resident Doctor, Department of Radio-diagnosis, JLN Medical College, Ajmer.⁴2nd Year Resident Doctor, Department of Radio-diagnosis, JLN Medical College, Ajmer.**ABSTRACT**

Takayasu's arteritis (Idiopathic Medial Arthropathy) or "Pulseless" disease is a rare, idiopathic, chronic granulomatous vasculitis that affects aorta and its major branches. This report describes a 30-year-old female who presented with generalised weakness, fatigue, headache, and bilateral arm claudication. Her bilateral upper limb colour duplex study revealed bilateral common carotid arteries and subclavian arteries luminal narrowing and CT angiography revealed vessel wall thickening of aorta and its branches with occlusion. She was diagnosed as having Takayasu's disease (Type V).

KEYWORDS

Takayasu Arteritis (TA), Colour Doppler Study and CT Angiography (CTA).

HOW TO CITE THIS ARTICLE: Gupta A, Bagoria D, Pamnani J, et al. Case report of Takayasu's arteritis. J. Evid. Based Med. Healthc. 2016; 3(44), 2232-2235. DOI: 10.18410/jebmh/2016/494

INTRODUCTION: Takayasu's arteritis is a chronic, idiopathic, inflammatory disease that primarily affects large vessels, such as the aorta and its major branches, pulmonary and coronary arteries.^[1] The most frequently affected arteries being the subclavian (90%), carotid (45%), vertebral (25%), and renal (20%). It occurs most frequently in young Asian women, although the disorder has been observed worldwide. Stenosis and obstruction are characteristic of the disorder, but dilatation and aneurysms are not rare. Sonography in conjunction with colour and pulsed Doppler imaging is a valuable tool in the evaluation of luminal narrowing of vessels in aortoarteritis. We can accurately diagnose, grade, and follow the progress of the disease. Conventional angiography has been traditionally considered the gold standard for the diagnosis of TA.

However, MDCT angiography is emerging as a reliable tool in non-invasively depicting both luminal and mural lesions in the aorta and its main branches, which may facilitate the detection of vasculitis during the early phase of TA. In late-phase TA, angiography usually demonstrates luminal changes such as stenosis, occlusion, or aneurysmal dilatation of the aorta and pulmonary artery and their branches. However, absence of such luminal changes does not exclude the possibility of early-phase TA. Familiarity with the varied chest radiographic, angiographic, CT, and MR imaging features of Takayasu arteritis will permit earlier diagnosis and treatment. In this case report, we will review the colour Doppler and CTA findings in TA.

CASE REPORT: A 30-year-old, married female had generalised body weakness, easy fatigability of upper extremities, cold fingers, numbness and pain in the upper extremities that worsened on exertion resembling ischaemic character along with significant history of weight loss, headache and anaemia for 3 years. Her cardiac risk profile was negative for diabetes mellitus, hypertension, and hyperlipidaemia. She had history of two uneventful deliveries.

Family history was not significant. On examination, she had diminished radial pulse and brachial pulse in both upper limbs. Her blood pressure was 90/60 mmHg in both upper limbs and 120/80 mmHg in both lower limbs. Cardiovascular examination revealed bruits in bilateral CCA and subclavian arteries. Cardiac auscultation was unremarkable. Peripheral pulses in both lower limbs were normal. Laboratory findings showed Increased ESR and positive CRP test. Echocardiography revealed grade 2 LVDD, mild AR, mild TR, mild PAH. Abdomen USG was normal. We further assessed patient in our department with colour Doppler study and CT angiography of both upper limbs which revealed following findings:

Colour Doppler:

- Right proximal subclavian artery showing marked circumferential thickening (70% narrowing).
- Left subclavian artery in its proximal part showing marked thickening and luminal narrowing by 72%.
- Distally blood flow in both upper limbs was of low resistance, mono-phasic up till radial and ulnar arteries suggesting ischaemic pattern.
- B/L CCA also show marked thickening and luminal narrowing by about 80-90%.
- Right ICA and ECA were narrow in calibre.
- Left ICA and ECA were normal in calibre.

Financial or Other, Competing Interest: None.

Submission 04-05-2016, Peer Review 19-05-2016,

Acceptance 26-05-2016, Published 02-06-2016.

Corresponding Author:

Dr. Divya Bagoria,

2nd Year Resident Doctor, Department of Radio-diagnosis,

JLN Medical College, Ajmer.

E-mail: divya.bagoria11@gmail.com

DOI: 10.18410/jebmh/2016/494

CT ANGIOGRAPHY OF UPPER LIMBS: There was evidence of thickening of the wall of entire aorta (Arch, Ascending, and Suprarenal Renal Descending Aorta).

Evidence of wall thickening with imperceptible enhancement of bilateral CCA, left CCA showing approximately 90% stenosis but ipsilateral ICA & ECA showing normal enhancement with mild wall thickening of vessels as they are patent from collaterals from respective vertebral arteries. Bilateral subclavian arteries showing 72% narrowing with collateral formation at parascapular and posterior intercostal region. Above findings were suggestive of type 5 TA. Vessel wall calcification was not present.

DISCUSSION: TA 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 ACR criteria 1990 (Sensitivity 77.4%, specificity 95%) considers three of the following six criteria necessary for a definite diagnosis of TA:

1. Age at onset \leq 40 yrs.
2. Claudication of extremities while in use.
3. Decreased brachial artery pulse.
4. Difference of Systolic BP $>$ 10 mmHg between arms.
5. Bruit over arteries.
6. Arteriographic abnormality: narrowing or occlusion of the entire aorta or its proximal branches.

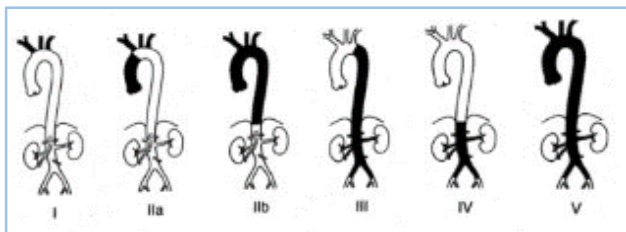


Fig. 1: New angiographic Classification of TA (6 types).^[2]

1. Type I involves only the branches of the aortic arch.
2. Type IIa involves ascending aorta, aortic arch and its branches.
3. Type IIb affects ascending aorta, aortic arch and its branches, and thoracic descending aorta.
4. Type III involves the descending thoracic aorta, the abdominal aorta and/or the renal arteries. The ascending aorta, the aortic arch and its branches are not affected.
5. Type IV involves only the abdominal aorta and/or renal arteries.
6. Type V=Type IIb + IV (Most Common Type).

Additionally, involvement of the coronary and pulmonary arteries should be indicated as C (+) or P (+), respectively.

Sonographic Findings: Sonography is the primary investigation of choice. Findings can be following according to the nature of the lesion.

1. Wall Thickening:

- Earliest finding in aortoarteritis and is universally seen in all patients.
- Uniform thickening of the wall of the vessels involved.
- The earliest wall thickening is seen in the SCAs, most commonly the left SCA. The AoA is also involved early.
- In Takayasu's arteritis, long segments of diffuse, homogeneous, moderately echoic circumferential vessel wall thickening are found. This is seen more commonly in the CCA and has been described as the "Macaroni Sign".
- It can be distinguished arteriosclerosis, which is more inhomogeneous. An increase in wall thickness is associated with secondary signs such as decreased pulsatility and loss of a normal triphasic flow pattern. The involved vessels reveal loss of the triphasic pattern, with a monophasic or biphasic parvus tardus type of spectral flow pattern. This type of pattern is also seen distal to an occlusion when there is reformation of vessels by collaterals, but wall thickening associated with dampened flow suggests the diagnosis of aortoarteritis.

2. Luminal Stenosis/Narrowing:

- It is commonly seen as a long segment compared with atherosclerosis or fibromuscular dysplasia, in which the stenoses are commonly short segments.

3. Luminal Dilatation and Aneurysms:

- Not as common as narrowing. Due to inadequate supportive fibrous tissue or focal intima weakness. The aorta is most commonly
- Affected, especially the thoracic and abdominal portions.

4. Calcification:

- Uncommon in aortoarteritis and more commonly seen in atherosclerosis.

5. Occlusions:

- Seen in later stages of the disease.
- Smaller vessels such as the carotid, subclavian, vertebral, and renal arteries are commonly involved. Because of the chronicity and slow progression of the disease, occlusions are commonly associated with collateral flow.

6. Pulsatility/Compliance:

- It has been found to be decreased in the pulseless stage of the disease in all cases of aortoarteritis.
- Changes in compliance have been known to precede the angiographic changes in vessels affect by aortoarteritis.
- Measurement of arterial compliance may provide indices of early vascular changes that predispose to the development of major vascular disease.

CT Angiography Features:

1. Mural thickening: Acute active phase.^[3]

- The typical manifestation of TA is the concentric mural thickening of the involved arteries.^{[3],[4]} Calcification in the thickened wall is another important sign.
- It is usually transmural and has been observed in 27% of patients
- On pre-contrast CT scanning, the mural thickening is of high attenuation compared with the lumen, while on the post-enhanced CTA images, it exhibits a double ring enhancement pattern, which is typically shown in venous phase. Specifically, a poorly enhanced inside ring and an obviously enhanced outside ring is frequently observed. It has been proposed that the inside ring represents the swollen intima, while the outside ring indicates the active inflammation in the medial and adventitial layers.

2. Luminal Changes:

- Stenosis is the most commonly seen finding associated with mural thickening, and observed in approximately 90% of patients.^{[4],[5]}
- Luminal stenosis of the abdominal and thoracic descending aorta has been reported in more than 60% of patients.
- Occlusion, ectasia and aneurysm of the vessels can also be seen, but less commonly. Dilatation and aneurysms are usually seen in the ascending and abdominal aorta, respectively, which may lead to fatal consequence, such as aortic rupture.

3. Collateral Formation

- It is observed in some cases and helpful in planning and modifying treatment.
- Maximum intensity projection and VR are useful in demonstrating small vessel changes.

4. Other Findings

- The pulmonary arteries are also commonly involved, with the most common appearance being peripheral pruning. Pulmonary and coronary artery involvement and ischaemia of the end organs such as decreased perfusion in the brain and lung can also be demonstrated on CTA images which is helpful in evaluating prognosis.^[6]

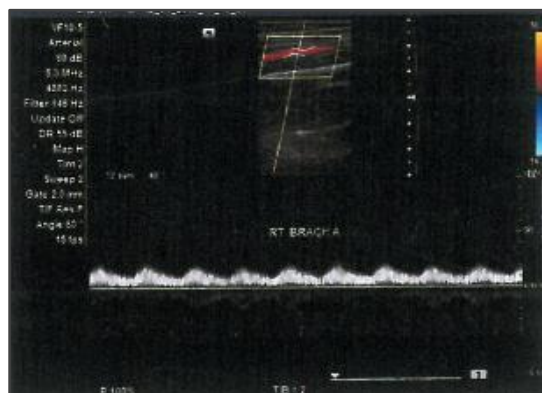
Differential Diagnoses:

- **Atherosclerosis:** It is not easy to differentiate aortic calcification in TA from that in atherosclerosis. It is more common in patients aged >45 years, and usually associated with short segment luminal stenosis. Calcification in ascending aorta can be observed in some TA patients, but it is rare in atherosclerosis.

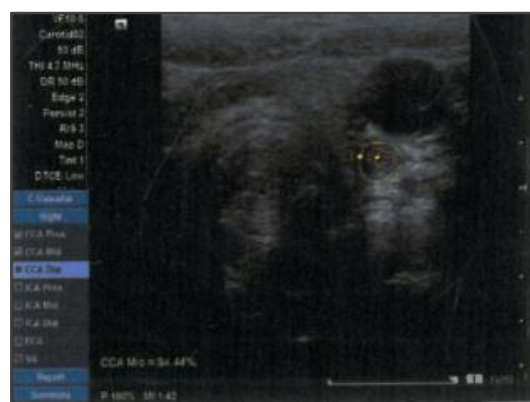
- **Giant Cell Arteritis:** It commonly affects patients >50 years and branches of the external and internal carotid arteries are most frequently diseased.
- **PAN:** It frequently occurs in adults who are 30–50 years old, affecting males more than females, and more commonly affects patients with hepatitis B. Gastrointestinal and renal arteries are most commonly involved. Multiple small aneurysm formation in the involved artery is the characteristic manifestation on CTA images.

Treatment:

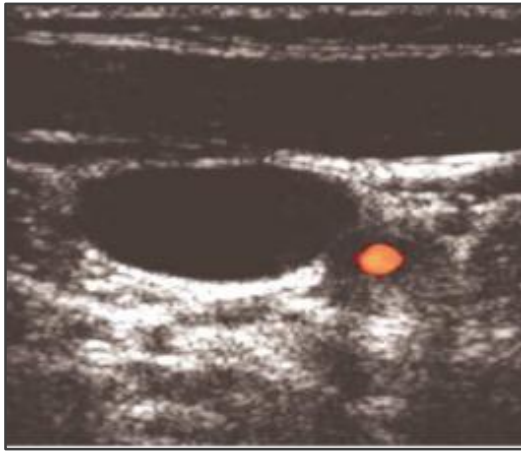
- Corticosteroid is mainstay of treatment. In patients with symptomatic stenotic or occlusive lesions, percutaneous transluminal angioplasty and stenting or bypass surgery is the most common palliative treatment.
- Following surgery or percutaneous angioplasty with or without endovascular stenting, long-term follow-up should be initiated with CTA or MRA to monitor the patency and complications of diseased arteries, stent or bypass vessels.



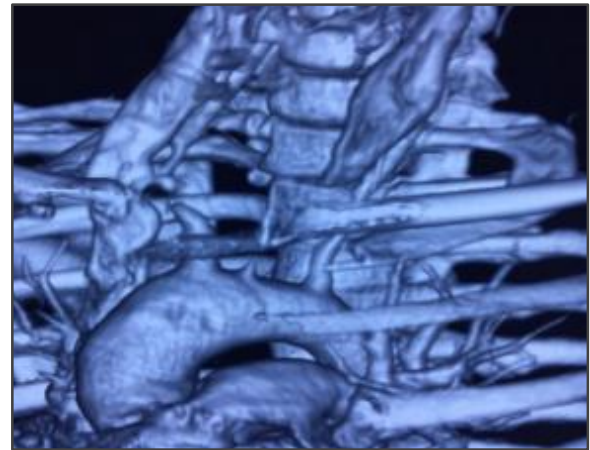
On Spectral Doppler Rt Brachial Artery Shows Tardus Parvus Waveform Distal To Stenosis Of Ipsilateral Subclavian Artery



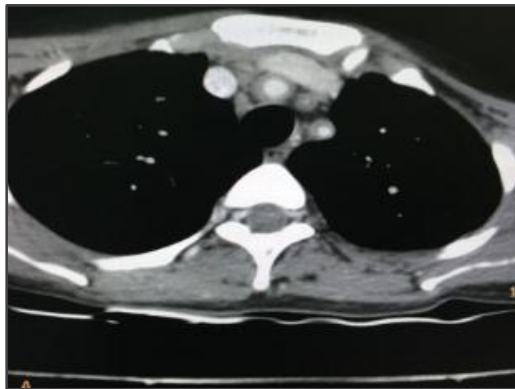
On Grey Scale Left CCA Shows Circumferential Narrowing Causing Luminal Stenosis of Approx .94%



On Colour Doppler Left CCA shows MACARONI Sign Due to Circumferential Luminal Narrowing



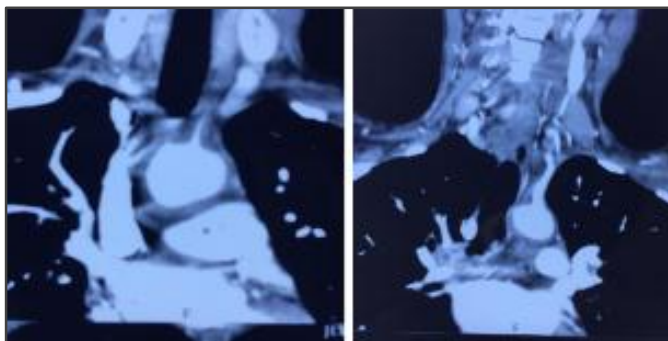
Coronal 3D Reformatted Image Shows Narrowing of AoA and its Major Branches



Ct Angiography (Axial Cut) Shows Circumferential Wall Thickening of Lt CCA, Lt SCA and Rt Brachiocephalic Artery



CT Angiography Shows Almost Complete Non-Opacification of Bilateral CCA



Coronal CT Angio Shows Luminal Narrowing of Left Subclavian Artery, Left CCA & AoA

REFERENCES

1. Mason JC. TA: advances in diagnosis and management. *Nat Rev Rheumatol* 2010;6:406-415.
2. Hata A, Noda M, Moriwaki R, et al. Angiographic findings of TA: new classification. *Int J Cardiol* 1996;54:S155-163.
3. Khandelwal N, Naveen Kalra, Mandeep Kumar Garg, et al. MDCT angiography in TA. *Eur J Radiol* 2011;77(2):369-374.
4. Canyigit M, Peynircioglu B, Hazirolan T, et al. Imaging characteristics of TA. *Cardiovasc Intervent Radiol* 2007;30(4):711-718.
5. Zhang LJ, Lu GM. TA involving the pulmonary arteries: evaluation by quantitative dual-energy CTPA. *Eur Heart J* 2012;33(7):928.
6. Katz-Agranov N, Tanay A, Bachar DJ, et al. What to do when the diagnosis of giant cell arteritis and Takayasu's arteritis overlap. *Isr Med Assoc J* 2015;17(2):123-125.