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Takayasu Arteritis-Evaluated by CT Angiography

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Abstract: ***Takayasu arteritis is a rare, chronic, idiopathic, granulomatous, inflammatory disease primarily involving large vessels like aorta and its major branches and pulmonary artery but can also involve coronary arteries. Also known as idiopathic medial aortopathy or pulse less disease. There is strong female predominance (F:M :: 9:1) and it tends to affect younger patients (<50 years of age). Here is a case of 37 years old female patient with complaint of headache and left sided upper limb weakness. Diagnosis of Takayasu arteritis made by CT angiography.***

Keywords: Takayasu arteritis (TK), CT angiography (CTA), Aorta, Vasculitis, Pulseless

1. Clinical History

Here we are presenting a case of 37 year old female presenting to medicine department of our hospital with the complaint of headache and left sided upper limb weakness. The patient had headache since 3 months which was gradually increasing in frequency. There was no associated vomiting, fever, convulsion or chest pain .On local examination there were absent pulsations in left upper limb arteries with normal pulses in other extremities. The pulse was increased along with high blood pressure of 146/92mmHg.Her MRI Brain showed multiple small focal recent ischaemic areas in the cortical sub-cortical areas of brain parenchyma.2D-Echocardiography was performed for this patient which showed symmetrical thickening of the aorta. The ejection fraction was normal.

2. Imaging Findings

On CT angiography circumferential symmetrical wall thickening was noted involving ascending aorta, arch of aorta and its branches along with descending aorta upto the D7vertebrae. The wall thickening caused mild luminal narrowing of branches of arch of aorta. Involved branches of arch of aorta-brachiocephalic artery, proximal part of bilateral common carotid and right subclavian artery and left subclavian artery upto inner border of 1st rib shows severe luminal narrowing with distally opacified by collaterals. The intimomedial thickness of ascending aorta, arch of aorta and descending aorta was 7.5mm,5.5mm and 4.8mm respectively. The patient was diagnosed with Takayasu arteritis type IIb.

3. Discussion

3.1 Background

Takayasu Arteritis is a rare, chronic, progressive, inflammatory, granulomatous, large vessel vasculitis commonly occurring in middle aged females. Most of the patients between the age of 20-40 years.

Arteries involved are aortic arch and its branches, pulmonary arteries, renal arteries, coronary arteries.

Carotid stroke/ Transient ischemic attack (TIA) is frequent in Takayasu arteritis.

Clinically arteritis can be classified into active inflammatory phase-pre-pulseless phase and chronic phase-pulseless phase. The active phase is characterised by systemic diseases with signs of fever, malaise, night sweats, weight loss and arthralgia lasting for weeks to months. In late chronic phase symptoms comprise of absent radial pulses in left upper limb, hypertension, hypertensive retinopathy, LV dysfunction, peripheral leg ulcers and congestive heart failure.

According to American College of Rheumatology, at least three out of the following six criteria should be satisfied for a definite diagnosis of Takayasu Arteritis. These criteria include, age under 40 at disease onset, claudication of extremities, decreased brachial artery pulse, blood pressure difference more than 10mm Hg between arms, a bruit over subclavian arteries or aorta and angiogram abnormalities: occlusion or narrowing in the aorta or its main branches.

The patient also had raised ESR, CRP reflecting the underlying inflammatory process. The laboratory investigations are nonspecific for the diagnosis of TA. There is no serological test for TA and the symptoms are constitutional leading to difficulty in early diagnosis.

3.2 Pathophysiology

Takayasu arteritis was characterized as an inflammatory granulomatous vasculitis of medium and large arteries, which leads to transmural fibrous thickening of the arterial walls, leading to multiple vascular obstructions and eventual ischemic changes. Degeneration of elastic fibers with the formation of aneurysms occurring when inflammation leads to loss of medial smooth muscle cells.

3.3 Role of Ctangiography

Conventional angiography has been traditionally considered the gold standard for the diagnosis of TA. However, multidetector CT angiography (CTA) 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. CTA will allow for visualization of vessel wall thickening and luminal narrowing [1]-[2}.

CT angiography depicted mural changes, including wall thickening, calcification and mural thrombi not seen with conventional angiography. The sensitivity and specificity of CT angiography in the diagnosis of Takayasu arteritis were 95% and 100%, respectively [3].

The typical manifestation for TA on CT images is the concentric mural thickening of the involved arteries may be the most important finding in the early phases of the disease. Transmural Calcification in the thickened wall is another important sign of TA. 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 (a poorly enhanced inside ring-the swollen intima and an obviously enhanced outside ring-active inflammation in the medial and adventitial layers) which is typically shown in venous phase [1].

Curved planar reformation (CPR) allows tortuous vessels to be displayed along its long axis; multiplanar reconstruction (MPR) gives the anatomical information of arteries in the optimal planes; volume-rendered (VR) images can illustrate the extension of the luminal lesions and map the collaterals following artery occlusion. A combination of CPR, MPR, VR, and axial images permits optimal evaluation of luminal changes. Sometimes, the normal or dilated proximal vessels associated with tapered narrowing of distal segments exhibit a characteristic “rat tail”-like configuration[1].

3.4 New Angiographic Classification of Takayasu Arteritis:

The classification of the disease is based on the involvement of blood vessels into different types[5].

Type	Vessel Involvement
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

The involvement of the coronary or pulmonary arteries is designated by C(+) or P(+)
Our patient was diagnosed as TA Type IIb as there was concentric wall thickening of ascending aorta, aortic arch and its branches, thoracic descending aorta.

4. Conclusion

Computed tomography angiography (CTA) performed with a 256-slice unit revealed high effectiveness in localization of vascular wall and lumen pathologies resulting from Takayasu arteritis. It is now possible to perform successful monitoring of patients with Takayasu arteritis and to plan possible interventional treatment. Multi-slice CTA allows for quick and precise diagnostic imaging in vascular diseases, enables accurate follow-up of vascular wall morphology and width of vascular canal regardless of localization of lesions.

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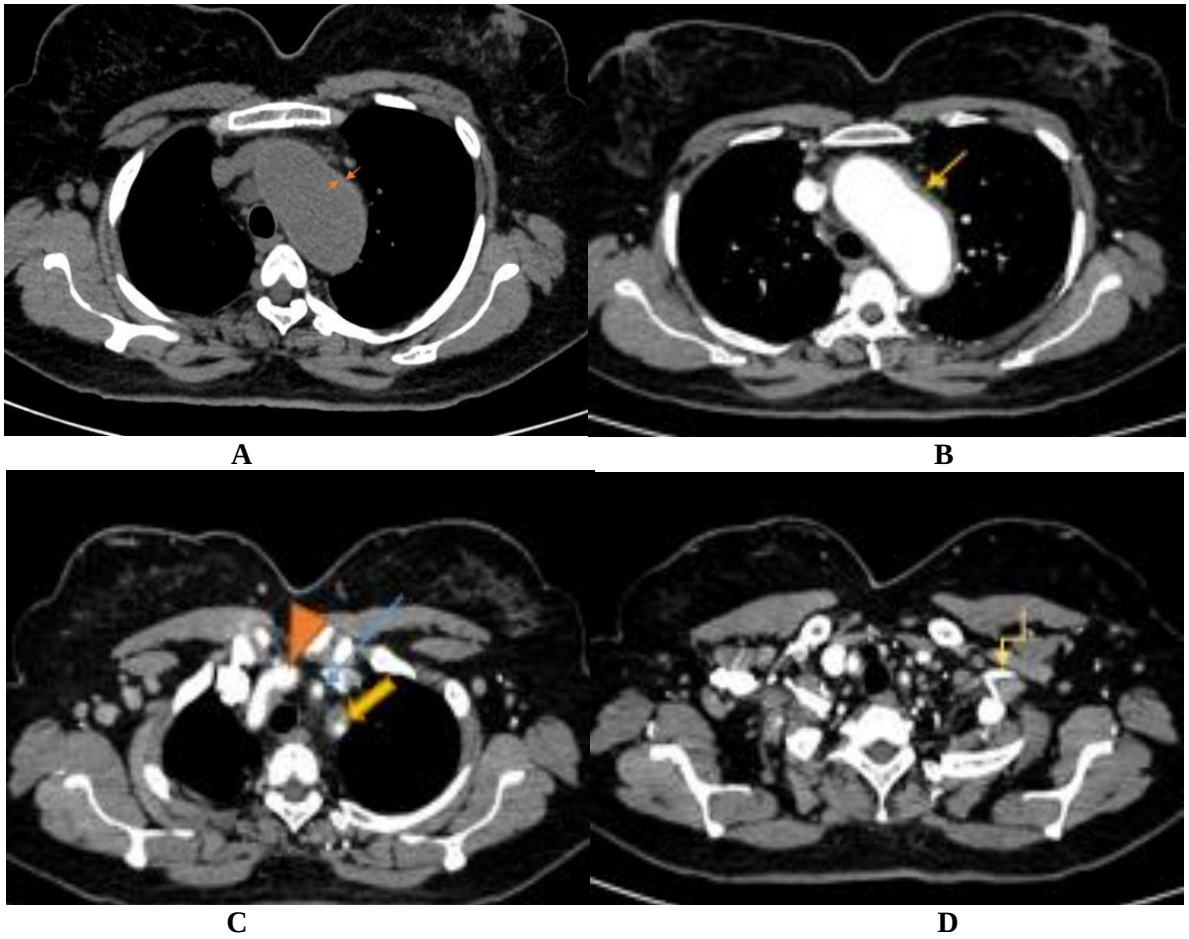
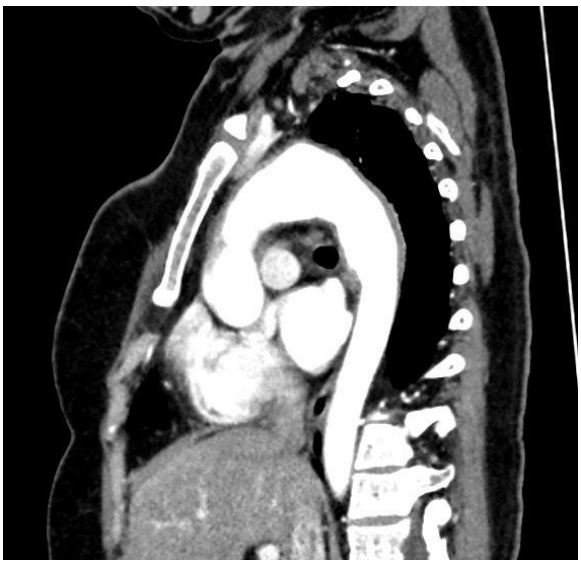
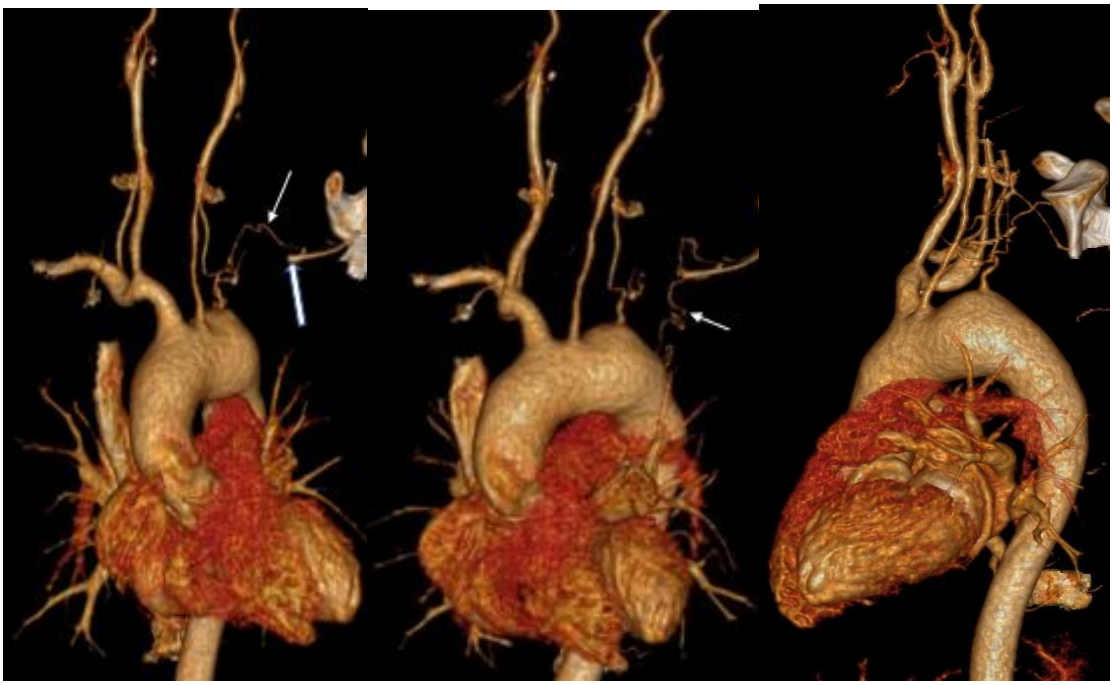


(A)Axial pre-contrast CT image shows the concentric thickened high-attenuation wall [arrows in A] in arch of aorta. (B) the wall appears low-attenuation compared with the lumen [arrow in (b)] on post-contrast images. (C) brachiocephalic (arrowhead), left common carotid artery (large arrow) and left subclavian artery (block arrow) are involved. (D) left subclavian artery distally opacified by collaterals (curved arrow)



Oblique rendered image shows the concentric thickened low-attenuated wall of ascending aorta, arch of aorta and descending aorta on post-contrast image.



Volume-rendered reformatted image shows the luminal stenosis of the diseased arteries. With left subclavian artery (block arrow)distally opacified by collaterals (arrow).