

Takayasu Arteritis Diagnosed by Carotid artery Doppler: A Rare Case Report

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Abstract

Takayasu arteritis is a rare, granulomatous large vessel vasculitis which is also known as “Pulseless disease” or “Martorell syndrome”. Chronic inflammation leads to stenosis, occlusion and aneurysm of vessels. Here we present a case of a 40 years old male patient diagnosed as Type-I Takayasu arteritis using carotid doppler and confirmed by CT angiography.

Introduction

Takayasu arteritis is a rare large vessel vasculitis first described in 1908 by a Japanese ophthalmologist, Mikito Takayasu. Chronic granulomatous inflammatory changes progress to stenosis, occlusion and aneurysm of medium and large arteries¹, mostly affecting the aorta and its branches. Incidence of the disease is higher in Asia with a strong female predominance (F:M-9:1).² Patients are usually younger than 50 years.² Early clinical diagnosis is challenging due to non-specific symptoms and rarity, where imaging can play a vital role. Now a days, USG, CT scan, MRI and PET are available. Although angiography is the gold standard for diagnosis, color doppler study is the most available, economic, non-invasive, non-ionizing tool for the assessment of the vessels.^{3,4}

Case Report

A 40 years old male patient was admitted to Sir Salimullah Medical college & Mitford Hospital (SSMC&MH) on 5th January, 2023 with the complaints of right sided weakness and headache. He had a history of fall around 08 days back followed by loss of consciousness. He was non diabetic, non-asthmatic & gave no previous history of hypertension. On admission he was non anemic,

non-icteric, GCS-11/15, Heart rate-85 BPM & respiratory rate-17BR/min. Peripheral pulse was not palpable & blood pressure was not recordable.

Investigations revealed increased ESR & positive C-reactive protein. CT scan of brain revealed large hypodense infarct at left temporoparietal region. ECG was normal. With the above findings the patient was referred to our center for doppler study of carotid and upper limb vessels.

Doppler study of carotid vessels was done with a high frequency (5-12 MHZ) linear transducer that revealed diffuse wall thickening and luminal narrowing in brachiocephalic trunk. Circumferential, diffuse, homogeneous wall thickening of the whole length of right common carotid artery including bulb along with marked luminal narrowing giving characteristic macaron sign resulting 70% diameter reduction and 90% area reduction. Circumferential wall thickening is also noted in the proximal part of the left common carotid artery. External and internal carotid arteries were unremarkable on both sides. Marked low PSV observed in all neck vessels. Doppler study of both upper limb vessels showed thickening of both subclavian arterial walls along with monophasic, dampened flow spectrum in all upper limb vessels. Doppler of renal artery and abdominal aorta showed no abnormality. After the doppler findings Takayasu arteritis was the leading consideration due to the presence of characteristic macaron sign. Then Patient was advised CT angiography for further confirmation. It revealed long segment narrowing of brachiocephalic trunk (about 27mm) and proximal part of right subclavian artery (about 16mm), severe narrowing of right common carotid artery, circumferential wall thickening at the origin of left common carotid artery (3mm) resulting 70%-80% luminal narrowing and short segment stenosis at the proximal part of left subclavian artery. Arch & descending thoracic aorta appeared normal.

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Figure 1: USG of neck vessels longitudinal view (a) showed circumferential wall thickening of right common carotid artery (*) causing marked luminal narrowing & transverse view (b) showed diffuse, circumferential wall thickening giving macaron sign (*).

Finally, the patient was diagnosed as a case of Type-I Takayasu arteritis after clinical & laboratory findings & based on the vessels involved in imaging findings. After diagnosis oral prednisolone & methotrexate were started. Two weeks after oral steroids there was significant clinical improvement but doppler findings remained almost similar.

DISCUSSION

Takayasu arteritis is a rare, inflammatory large vessel vasculitis of unknown etiology that predominantly involves aorta and its branches, and may also involve pulmonary and renal arteries. Some studies show geographic concurrence of tuberculosis (TB) and Takayasu arteritis, that supports the hypothesis of TB triggered immune mediated reaction to vessel wall.²



Figure 2 (a & b): CT Angiogram of neck vessels confirmed involvement of aortic branches and common carotid arteries bilaterally

Three phases have been mentioned in different literature— early or pre pulseless phase followed by phase of active vascular inflammation & advance or pulseless phase. Early phase is characterized by non-specific symptoms like low grade fever, malaise, arthralgia, weight loss etc. Granulomatous inflammation of the vessel wall leads to fibrosis, stenosis, aneurysm and thrombosis characterized by ischemic manifestations in advance stage (2,3). Ischemic stroke was observed as initial presentation in 10-20% patients with Takayasu arteritis (1). Our patient also presented with ischemic stroke along with right sided hemiparesis. His pulse and blood pressure were non-recordable, reflecting that he was in a pulseless stage. Pulseless disease was named for reduced or absent peripheral pulses due

to obliterative changes in the aorta & its branches.²

Vascular imaging is the mainstay of diagnosis as clinical diagnosis is difficult in the pre pulseless stage due to non-specific symptoms. Imaging modalities reflects the wall thickening along with luminal narrowing as a result of inflammatory changes. Doppler sonography is the widely available, noninvasive technique to assess vessel wall thickening, luminal narrowing, stenotic and aneurysmal dilatation. Loss of normal triphasic flow as a result of decreased arterial pulsatility resulting in a tardus parvus spectrum which can be observed non-invasively.^{3,5} Homogeneous, diffuse, circumferential wall thickening of arteries in Takayasu arteritis produce macaroni sign.⁶ This decisive sign was observed in the right common carotid artery of our patient. Diffuse, homogeneous thickening differentiates it from atherosclerotic thickening which is irregular and more inhomogeneous.³

Angiography is the gold standard for diagnosis. Although digital subtraction angiography is more accurate for assessing vessel morphology, it is being used less frequently due to high radiation burden and invasiveness. CT angiography (CTA) has high sensitivity in assessing mural changes. Oura et al., suggests CTA is useful for diagnosis but not suitable for regular monitoring for radiation exposure. In our case, there was an excellent correlation between doppler & CTA findings and the patient was diagnosed as type I Takayasu arteritis non-invasively. MR angiography is another excellent vascular imaging modality but it is less accessible, time consuming and costly; however not useful for monitoring.^{1,7}

Early diagnosis is crucial for better prognosis and reduction of complications. Vascular imaging can detect the disease in the early phase when clinical diagnosis is difficult. Doppler sonography can be used for both diagnosis and monitoring due to its high accessibility, no radiation exposure, low cost and non-invasiveness in early and late phases^{5,6} Oura et al., suggested it is more subjective than

other vascular imaging modalities.¹

CONCLUSION

Takayasu arteritis is a rare entity among various types of vasculitis that can be diagnosed by clinical manifestations & radiological findings. This case report highlights the significance of doppler study as a reliable tool not only for diagnosis and grading but also for monitoring of the patient's response to treatment.

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