



IMAGING PEARLS OF A PULSELESS DISEASE – A REVIEW OF FOUR CASES OF TAKAYASU ARTERITIS

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ABSTRACT

Takayasu arteritis is an autoimmune disorder affecting the media of large arteries for which the exact cause is still unclear [1-3]. It usually affects the aorta and its proximal branches, usually from the aortic arch. When it does affect the aortic arch, it can involve the carotid, vertebral and subclavian branches. It is characterized histologically by granulomatous inflammation that causes thickening of the media and intima of major arteries [8-10]. The disease is usually diagnosed in the later stages when it becomes clinically apparent. It is hypothesized to be due to an autoimmune response to an antigen that is similar to tuberculin [1-3, 8]. Through the advances in imaging techniques and with better equipped sophisticated imaging equipment, early diagnosis of Takayasu arteritis is possible. The utility of ultrasound lies in the early diagnosis of Takayasu arteritis when vessels accessible by sonogram like carotid and subclavian arteries are involved [4-7]. Characteristic imaging appearance of the disease in ultrasound along with typical signs [8] has been found to be very useful in diagnosing the medial changes seen in the disease. In this context, we describe here four cases of Takayasu arteritis which were diagnosed with ultrasonogram.

KEYWORDS :**INTRODUCTION**

Takayasu arteritis is not an uncommon disorder especially in the Asian populace. It usually affects young people in the second to third decade of life. The disease is characterized by autoimmune mediated processes which have not been fully understood. It is thought to be due to a response to tuberculin antigen although it has not been conclusively proved. The autoimmune pathological process initiates a cascade which results in granulomatous type of inflammation [1-3]. This results in long segment circumferential smooth wall thickening involving the medial layer of major arteries like the aorta and its branches [4-7, 9, 10]. The early stage of the disease also called as the 'pre-pulseless' stage is characterized by non-specific symptoms like fever, malaise, lethargy, fatigue, myalgia, arthralgia, and weight loss. The duration of the early stage is variable, and it can last even up to several years in some cases with symptoms occurring on & off [7]. Clinical criteria used for the diagnosis of Takayasu arteritis are usually met only the later stage of the disease called as the pulseless stage.

This stage is characterized by stenosis, thrombosis and aneurysm formation of the involved vessels which manifests clinically based on the vascular territory involved [7-10]. Complications may manifest which may be due to carotid stenosis, aortitis, subclavian stenosis, aortic aneurysm or renovascular hypertension. Pseudoaneurysm is a common acquired vascular complication that can occur along with stenosis of arterial segments and thromboembolism. Both the early manifestations and complications can be well visualized and diagnosed on ultrasound [4-10]. In this article, we review four distinct cases of Takayasu arteritis that were diagnosed in a tertiary care institutional hospital in Tamilnadu, India.

Case Report**CASE 1**

A 16-year-old female came with the complaints of giddiness for the past one week. There was also history of fever two days back and two episodes of syncope one day back. The patient was evaluated for routine infectious disease protocol initially

and in view of the history of giddiness, two dimensional ultrasonogram of the carotid and vertebral arteries was performed as a routine evaluation. The examination revealed the presence of circumferential increase in wall thickness of both the common carotid arteries. The wall thickening was smooth and showed moderate increase in echogenicity compared to the normal uninvolved vessel wall. And this was associated with luminal narrowing.

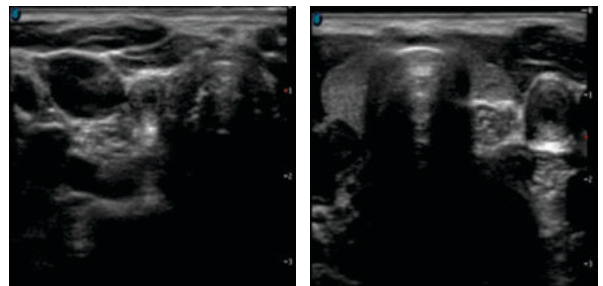


Figure 1-CASE 1: Axial view of the ultrasound of both common carotid arteries showing a circumferential increase in wall thickness that appears moderately echogenic, with luminal narrowing.

Longitudinal view of the vessels showed the presence of preserved flow with slightly high resistance in the involved arteries, with hemodynamic parameters being within normal limits.

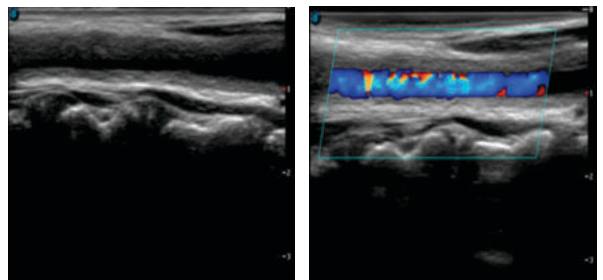


Figure 2-CASE 1: Longitudinal view of left carotid artery with preservation of colour flow.

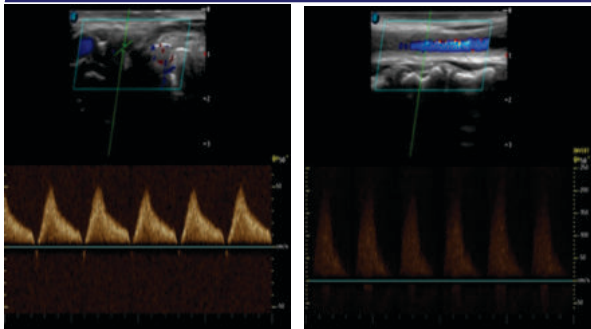


Figure 3 -CASE 1: Spectral doppler of both the carotid arteries shows slightly high resistance flow across the involved vessels with hemodynamic parameters being within normal limits.

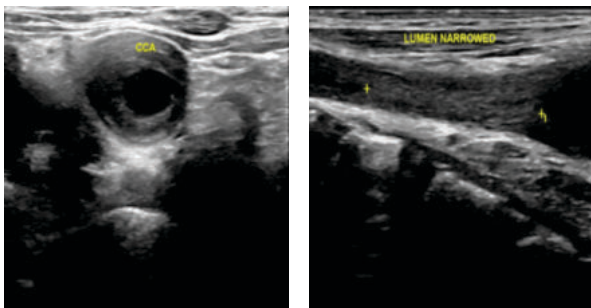


Figure 4 -CASE 1: Luminal narrowing of common carotid artery demonstrated in both axial and longitudinal sonographic views.

The patient was treated conservatively in the form of a steroid based regimen. The symptoms of the patient improved after starting the treatment and is on follow-up ever since.

CASE 2

A 20-year-old male came with complaints of headache and giddiness on and off for the past one year. Previously he was suspected of and was on medical treatment for vitamin deficiencies which did not improve his symptoms. 2D B mode and doppler ultrasound of the neck vessels was then done. Ultrasound showed the presence of echogenic circumferential increase in wall thickness with luminal narrowing of common, internal, and external carotid arteries on both sides.

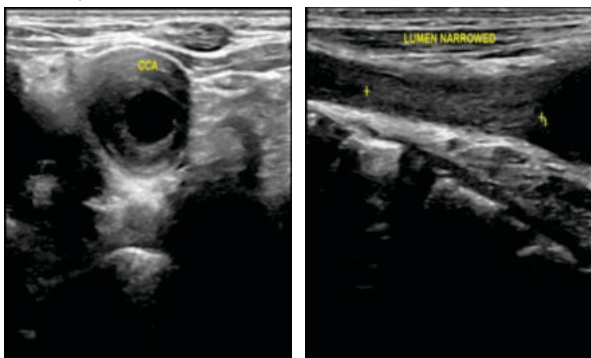


Figure 5 -CASE 2: B mode Ultrasound shows the presence of echogenic circumferential increase in wall thickness of the common carotid artery with luminal narrowing.

Further ultrasound examination showed a progressive increase in wall thickness in the distal common carotid artery in the right side that was associated with stenotic narrowing of the vessel.

The patient was started on steroid and other supportive medical treatment. There was a significant improvement in symptoms with treatment and the patient is on follow-up ever since.

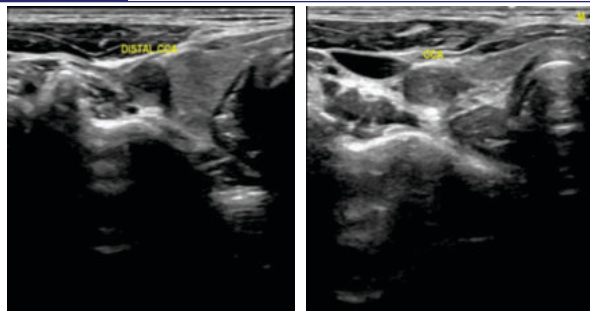


Figure 6-CASE 2: Ultrasound of the distal part of right common carotid artery shows progressive increase in wall thickness causing stenotic narrowing of the vessel.

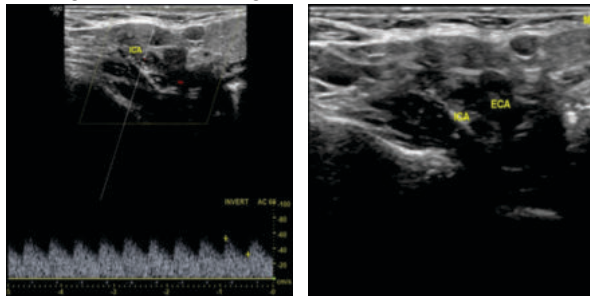


Figure 7-CASE 2: Ultrasound shows involvement of the internal and external carotid arteries with preservation of flow parameters.

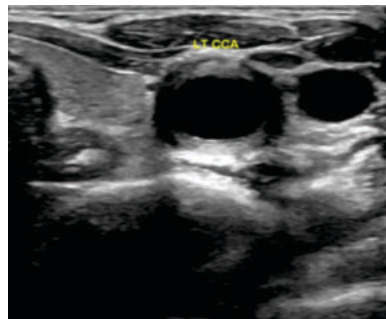


Figure 8 -CASE 2: Circumferential increase in arterial wall thickness giving the appearance of the characteristic 'macaroni' sign.

CASE 3

A 26-year-old male came with the complaints of pain and weakness of the right upper limb that developed gradually over the past one year. Patient was previously suspected of cervical rib causing vascular compression, but cervical radiograph had ruled out the diagnosis. Patient was also evaluated for possible young's stroke but magnetic resonance scanning of the brain was unremarkable. So, a B mode and doppler ultrasound of the carotid and subclavian system was performed. Ultrasound showed the presence of circumferential increased and echogenic wall thickening of the right subclavian artery. The patient improved with steroid treatment.

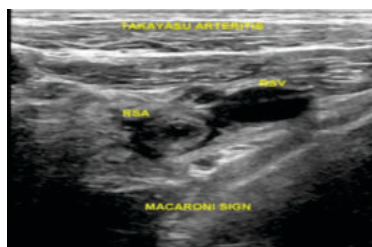


Figure 9 -CASE 3: Ultrasound showing concentric echogenic increase in wall thickness of right subclavian artery, the 'macaroni'sign.

CASE 4

A 14-year-old female patient came with the complaints of pain and mild weakness of right upper limb for the past 2 months. Magnetic resonance study of the brain of the patient done one month back was unremarkable. Ultrasound showed the presence of echogenic longitudinal concentric increased wall thickness of the arteries of the right upper limb with preservation of hemodynamic parameters.

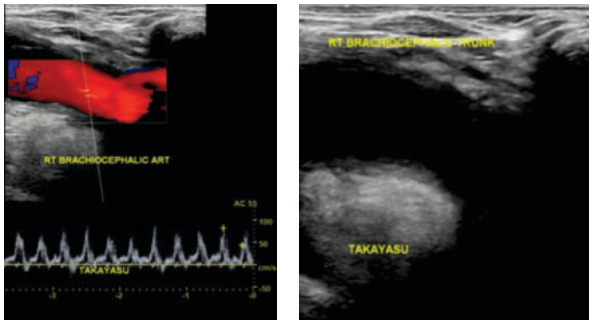


Figure 10 -CASE 4: Ultrasound shows involvement of right brachiocephalic trunk.

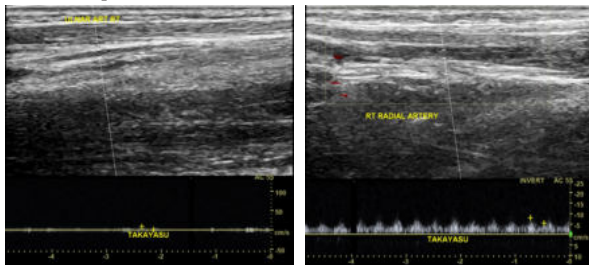


Figure 11-CASE 4: Ultrasound shows involvement of right radial and ulnar arteries.

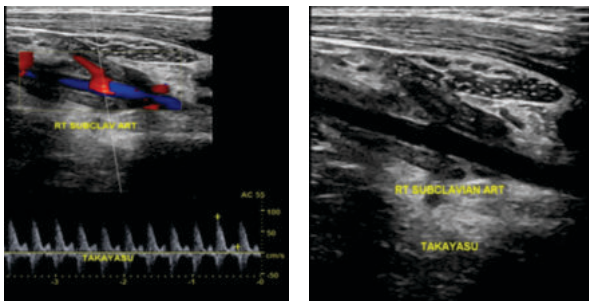


Figure 12-CASE 4: Ultrasound shows involvement of right subclavian artery.

There was involvement of right brachiocephalic trunk, right subclavian artery with involvement of distal arteries of the right upper limb namely the radial and ulnar arteries. The patient was provisionally diagnosed with Takayasu arteritis and started on immunosuppressive treatment. The patient's symptoms improved with treatment.

DISCUSSION

The etiopathogenesis of Takayasu arteritis may not be fully understood yet but available knowledge about the natural course of the disease allows to delineate the imaging findings at the early and advanced stages, and therefore helps to identify the disease where treatment or intervention can provide the maximum benefit [1-3, 9, 10]. The characteristic sonographic finding of a long segment circumferential wall thickening of the involved arteries which appears moderately echogenic compared to the normal medial wall of other vessel, helps to pinpoint the diagnosis at the early stage where complications have not yet set in. This finding is seen on axial views of the involved vessel as the characteristic 'macaroni' sign [8]. This utility of ultrasound can help to initiate early medical treatment that can potentially reverse the course of

the malady. It can also help to follow-up and prognostication of the disease course in patients who are under treatment [11]. Ultrasound imaging also helps as an initial screening modality in the identification of complications like thrombosis, which is seen as echogenic structure, which can be mobile or fixed. It can help to identify occluded or stenotic vessels. It can also help in the initial diagnosis of other complications like pseudoaneurysm, which occurs as a focal outpouching with a narrow neck on imaging [4-7]. Thus, ultrasound is an invaluable tool in the initial diagnosis and follow-up of Takayasu arteritis.

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