Review Article

Takayasu's arteritis : role of imaging

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Takayasu arteritis (TA) is a chronic large vessel vasculitis that affects aorta, its main branches and pulmonary arteries. The inflammatory process results in stenosis, occlusion, dilation or aneurysm formation in the arterial wall. TA has been described in different parts of the world and affects predominantly young individuals (<50 years of age). Patients with TA may present constitutional symptoms, vascular pain (e.g. carotidynia) and typical features such as limb claudication, decreased or absent peripheral pulses, vascular bruits, hypertension, and reduction or discrepancies in blood pressure between arms. A proper diagnosis of TA is an important issue since delays may result in significant morbidity. The definition of TA was included in the 1994 and 2012 Chapel Hill Consensus Conference and TA was categorized as a large vessel vasculitis. The first diagnostic criteria for TA were developed by Ishikawa in 1988 and modified by Sharma et al., in 1995. Different imaging modalities play a very vital role in the management of TA. Some of the modalities are useful in early diagnosis; some are helpful for monitoring disease activity and planning management. Current article aims at briefly discussing the role of different imaging modalities in TA.

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The first description of TA is usually attributed to Mikito Takayasu who presented a case of a 21 year-old woman with arteriovenous anastomosis surrounding the papilla on the eyeground in the 12th Annual Meeting of the Japan Ophthalmology Society in 1905. After Mikito Takayasu, the first reports of TA in medical literature in English were published in the early 1950s and in both publications TA was referred as "pulseless disease".

Takayasu arteritis (TA) is a chronic granulomatous large vessel vasculitis that affects the aorta, its main branches and pulmonary arteries[1]. The inflammatory process initially leads to thickening of the arterial wall and may result in stenosis, occlusion, dilatation or aneurysm formation. At disease presentation or during relapses, TA patients may present non-specific inflammatory complaints such as fever, malaise, anorexia, weight loss, myalgia or arthralgias which can be associated with vascular pain (eg carotidynia). As arterial lesions ensue, more characteristic features of TA may be found such as limb claudication, decreased or absent peripheral pulses, vascular bruits, hypertension, and reduction or discrepancies in blood pressure due to stenotic or occlusive lesions between arms. Heart failure may develop as a consequence of hypertension, coronary heart disease and/or aortic regurgitation. Transient ischemic attacks, stroke and mesenteric ischemia are other ischemic manifestations of TA. The vast majority of arterial lesions in TA are stenotic whereas aneurysms can be found in up to one third of TA patients[2]. Although geographical differences regarding the distribution of arterial lesions have been described in TA, the aorta is the most affected artery followed by subclavian, common carotid and renal arteries[3].

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Imaging in Takayasu's Arteritis :

In early-phase Takayasu arteritis, Computed Tomography (CT) and Magnetic Resonance (MR) imaging show thickening of the aortic wall. In late-phase Takayasu arteritis, angiography usually demonstrates luminal changes such as stenosis, occlusion, or aneurismal dilatation of the aorta and pulmonary artery and their branches[4,5]. However, absence of such luminal changes does not exclude the possibility of early-phase Takayasu arteritis. Familiarity with the varied chest radiographic, angiographic, CT, and MR Imaging features of Takayasu airteritis will permit earlier diagnosis and treatment.

Chest Radiographic Features :

The Chest radiographic manifestations include loss of sharp definition and a wavy or scalloped appearance of the descending thoracic aorta (Fig 1)[6]. Hayashi *et al*[7] reported that hilar enlargement, although rare, may be a new radio graphic finding in early-phase Takayasu Artentis (Fig 2). Such subtle findings in a young female patient should alert the radiologist to the diagnosis of early-phase Takayasu arteritis.

Ultrasonography :

Duplex Doppler ultrasonography (US) may be used to identify circumferential vessel wall thickening and thereby evaluate and monitor disease in the aorta and branch vessels (Fig 3). Sonography of carotids and subclavian arteries aids in detection of TA at an early stage, where characteristic long segment involvement with homogenous, midechoic, circumferential arterial wall thickening, which has been described as the "Macaroni's sign" may be found.[8] However, the examination is operator dependent and also depends on a suitable acoustic window. Furthermore, it has a low negative predictive value.

Angiographic Features :

Aortography may not show any intraluminal changes because the basic pathologic features of early-phase Takayasu Arteritis are mural changes in the great vessels[9]. On aortograms, the thickness of the wall of the descending thoracic aorta is measured as the distance between the intraluminal contrast medium and the air in the lung. This Measurement includes, the thickness of the two pleural layers (which may be negligible) and possibly the thickness should of periaortic infiltration. However, the thickness of the wall of the pulmonary artery is difficult to measure on angiograms.

Rarely, when the inflammatory changes are severe, granulomatous or diffuse productive in flammation in the media and adventitia is associated with marked secondary intimal hyperplasia, resulting in stenosis of the aortic lumen (Fig 4). Coronary involvement in TA is best delineated in conventional coronary angiography[10].

CT and MRI :

By demonstrating arterial wall changes, cross sectional imaging techniques such as CT and MR Imaging play an important role in early diagnosis of Takayasu artentis. The significant feature of early-phase Takayasu Artentis is aortic wall thickening.

Hayashi *et al* reported that a double ring pattern at enhanced CT is useful for early diagnosis of early-phase Takayasu Arteritis and evaluation of the effects of steroid therapy[7]. On unenhanced CT scans, the vascular wall is clearly distinguished from the vascular lumen by attenuation similar to or higher than that of muscle. On Enhanced CT scans, the wall shows the double ring pattern: a poorly enhanced in side ring and a well-enhanced outside ring (Fig 5). The inside ring is considered to represent mucoid or gelatinous swelling of the intima; the outside ring is considered to represent active medial and adventitial inflammatory Verse change.

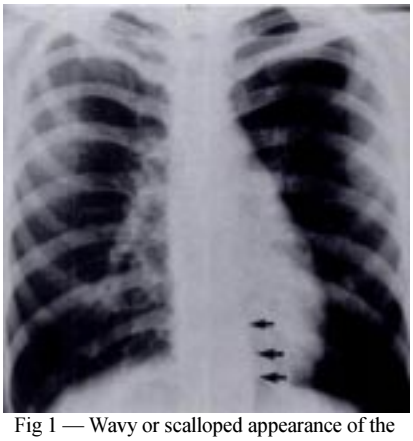

Fig 1 — Wavy or scalloped appearance of the descending thoracic aorta in chest radiograph

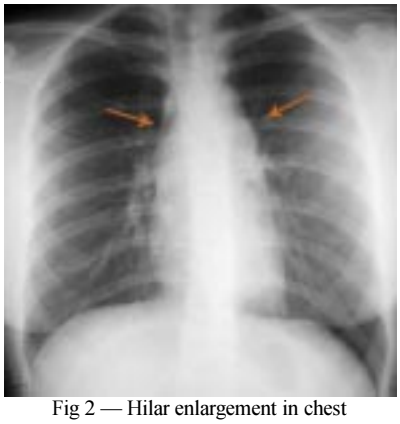

Fig 2 — Hilar enlargement in chest radiograph in a young female patient of Takayasu arteritis

The Inflamed arterial wall enhances at contrast-enhanced CT, because adventitial vascular structures in the aortic wall are probably enlarged vasa vasorum. In all 10 Healthy adults studied by Park et al, transverse arterial-phase spiral CT angiograms showed an aortic wall that was less than 1 mm thick or even imperceptible; the aortic wall could not be identified on precontrast and delayed images. Therefore, the sensitivity and specificity of CT for the detection of significant arterial wall thickening are thought

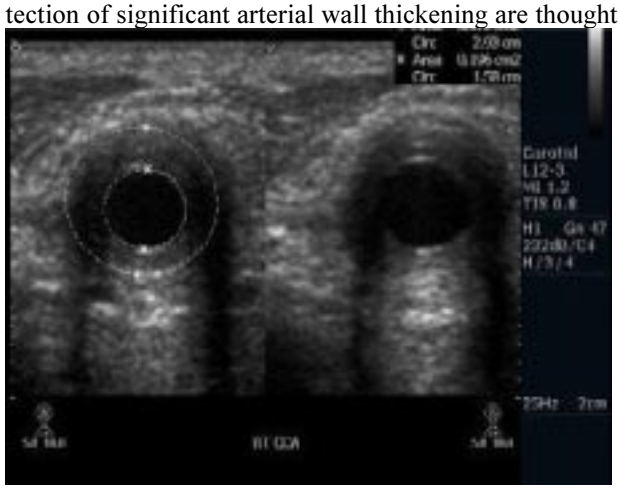

Fig 3 — Macaroni's sign in Sonography: homogenous, midechoic, circumferential arterial wall thickening in Takayasu arteritis

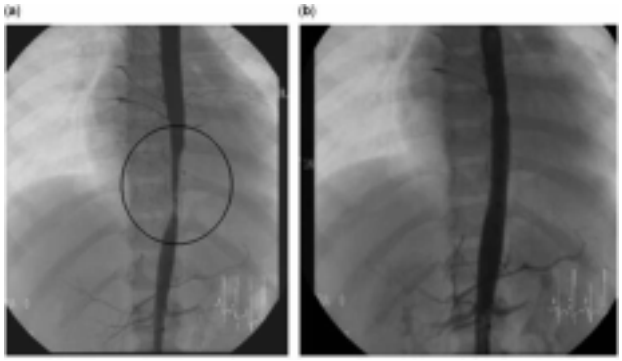

Fig 4 — Diffuse stenosis of abdominal aorta before (a) and after (b) stent angioplasty in a patient of Takayasu arteritis

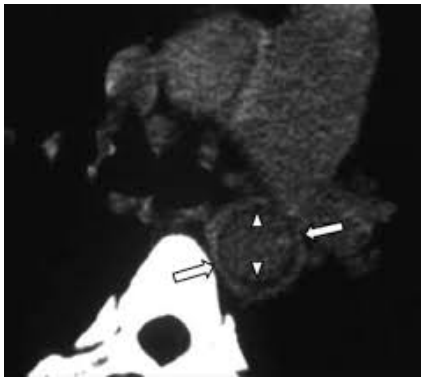

Fig 5 — Double ring pattern in CT Angiography: a poorly enhanced in side ring and a well-enhanced outside ring in a patient of Takayasu arteritis

to be high (Fig 5).

Arterial wall thickening can also be demonstrated with MR Imaging (Fig 6). MR imaging has the advantages of direct imaging in the axial, sagittal, and coronal planes with good contrast resolution, and the spin-echo technique allows differentiation of the arterial lumen from its wall without contrast medium[11]. An Imaging section perpendicular to the vessel of interest is best suited for assessing wall thickness with MR imaging. MR imaging in particular allows better soft-tissue differentiation and can show other signs of inflammation, including mural edema and increased mural vascularity. Other advantages of MR imaging are the lack of iodinated contrast material or ionizing radiation.

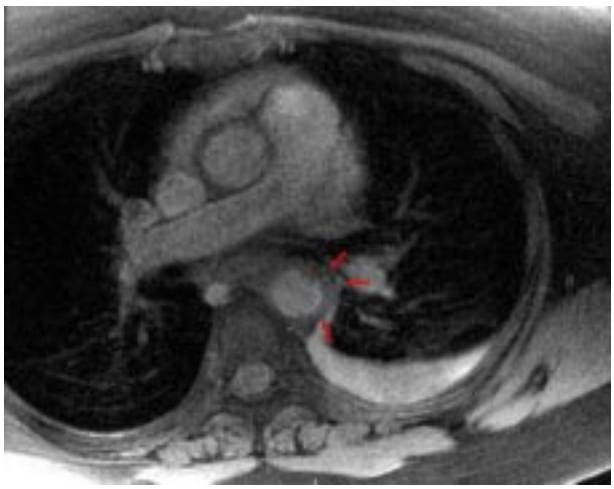

Fig 6 — Arterial wall thickening in a patient of Takayasu arteritis in MR Imaging

During the course of early-phase Takayasu Arteritis, either spontaneously or with steroid therapy, the aortic wall thickening may be reduced, corresponding to a decrease in active inflammation in and around the aortic wall. Steroid therapy has resulted in a dramatic improvement in clinical and radiologic abnormalities in patients with early-phase Takayasu Arteritis. Hayashi et al reported the first case of early-phase Takayasu Arteritis in which reduction of aortic wall thickening after steroid therapy was documented with CT[7]. Follow-up MR imaging of patients with early-phase Takayasu arteritis demonstrated significant reduction of wall thickening in the aorta and pulmonary artery after steroid therapy.

FDG-PET Scanning :

18F-fluorodeoxyglucose–positron emission tomography or FDG-PET scanning has been proposed as a new way of assessing disease activity in Takayasu arteritis (TA)[12]. This is a noninvasive metabolic imaging modality based on the regional distribution of 18F-fluorodeoxyglucose, which accumulates in hypermetabolic cells. Some authors have suggested that it could play a role in the management of large-vessel vasculitis because of its capacity to detect areas of increased glucose metabolism present in the vascular wall. Preliminary studies have shown that FDG-PET scanning had a sensitivity and specificity of 92% and 100%, respectively, for the assessment of active TA.

Conclusion :

Both conventional and modern imaging modalities play very vital role in the management of TA. These include early diagnosis, planning therapy, monitoring disease activity and also follow up of patients. Proper selection of imaging modalities as well as good interdepartmental coordination between rheumatologist, radiologist, physician and cardiologist is quintessential in formulating a perfect management plan.

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(Continued on page 49)

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