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INVITED REVIEW

CT and MR Angiography of Aorta and Aortic Branches in Giant Cell Arteritis

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Giant cell arteritis (GCA) is a granulomatous systemic vasculitis of large and medium-sized arteries predominantly affecting patients older than 50 years [1]. The external cranial arteries, such as the temporal artery, are commonly involved in GCA (cranial GCA). However, with the advances in imaging technology and more widespread use of cross-sectional imaging, frequent involvement of the aorta and its branches were also seen and referred to as extracranial large-vessel GCA (LV-GCA) [2,3].

While temporal artery ultrasound (US) and biopsy are commonly used as diagnostic methods, GCA may present without temporal artery involvement. Schmidt et al. reported negative US and histology findings of temporal arteries in 38% and 33% of LV-GCA patients, respectively, [4]. Also, in extracranial LV-GCA, the clinical presentation is often more subtle than temporal arteritis, ranging from non-specific constitutional symptoms to extremity claudication depending on the affected vessels [3,5]. Based on these, imaging techniques such as computed tomography (CT), magnetic resonance imaging (MRI), and 18F-fluorodeoxyglucose positron-emission tomography/computed tomography (FDG-PET/CT) have been increasingly used in the diagnosis of LV-GCA.

Using imaging, aortic involvement is reported in 45-65% of GCA patients, and the most commonly affected part of the aorta is the thoracic aorta (aortic arch and descending aorta) [6,7]. If abdominal aorta involvement is present, the thoracic aorta is also usually affected. Apart from the aorta, the supra-aortic branches- particularly subclavian and axillary arteries- are frequently involved. Carotid and vertebral artery involvement may be less often observed. Less commonly, visceral branches of the abdominal aorta and iliofemoral arteries may be affected [5,7–9].

To standardize imaging in large vessel vasculitis, European League against Rheumatism (EULAR) published some recommendations in 2018. According to these, early imaging before the treatment or as soon as possible after the therapy initiation is recommended to support the clinical and laboratory criteria in suspected GCA cases [3]. Because therapy with glucocorticoids reduces the sensitivity of the imaging [10], if there is a high clinical probability, the diagnosis of GCA can be made with positive imaging findings, eliminating the need for further investigations such as temporal artery biopsy. While in cranial GCA, first-line imaging should be temporal and axillary artery Doppler ultrasound, in extracranial LV-GCA, ultrasound, CT, MRI, or FDG-PET/CT is recommended to support the diagnosis. Conventional angiography is no longer recommended for diagnosis, considering significant disadvantages such as invasiveness, procedural risks, and the disability to show the vessel wall changes [3]. On the other hand, CT and MR angiography allows the evaluation of the aorta and its branches in a larger area in a single acquisition and the simultaneous assessment of the vessel wall and lumen.

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Note:
This manuscript was reviewed by Muşturay Karçaaltıncaba.

Therefore, the use of CT and MR angiography gained importance to address the involvement of the aorta and its main branches for diagnosis and disease monitoring in LV-GCA. Moreover, since GCA is seen in the elderly, findings of atherosclerosis such as wall thickening may interfere with imaging findings of large vessel vasculitis [11]. This review aims to describe CT and MR angiography findings of the aorta and its main branches' involvement in LV-GCA and focus on differentiating LV-GCA from atherosclerosis by imaging findings.

CT angiography in LV-GCA

CT angiography (CTA) is widely used in diagnosing large vessel vasculitis with a shorter acquisition time and superior spatial resolution. Multislice CT should obtain CTA with thin slices reconstructed with a thickness of 0.5-1 mm [12]. Besides the arterial phase, venous phase imaging should be performed to assess wall enhancement. The major disadvantage of CT is ionizing radiation exposure. Although the ongoing advances in radiation reduction algorithms partly overcome this, it is still a significant consideration in repetitive imaging during the follow-up [3,13,14].

The main imaging findings of LV-GCA are circumferential wall thickening and vessel wall enhancement depicted in the delayed/venous phase. Berthod et al. reported a wall thickness threshold of 2.2 mm, presenting the best sensitivity and specificity combination for GCA [15]. The wall enhancement on the venous phase exhibits a "double-ring" appearance characterized by a hypodense inner ring representing edematous intima surrounded by the hyperdense enhancing outer ring (Figure 1). Also, stenosis, occlusion, dilatation, and aneurysms may be observed in affected vessels [7,13,16,17] (Figure 2).

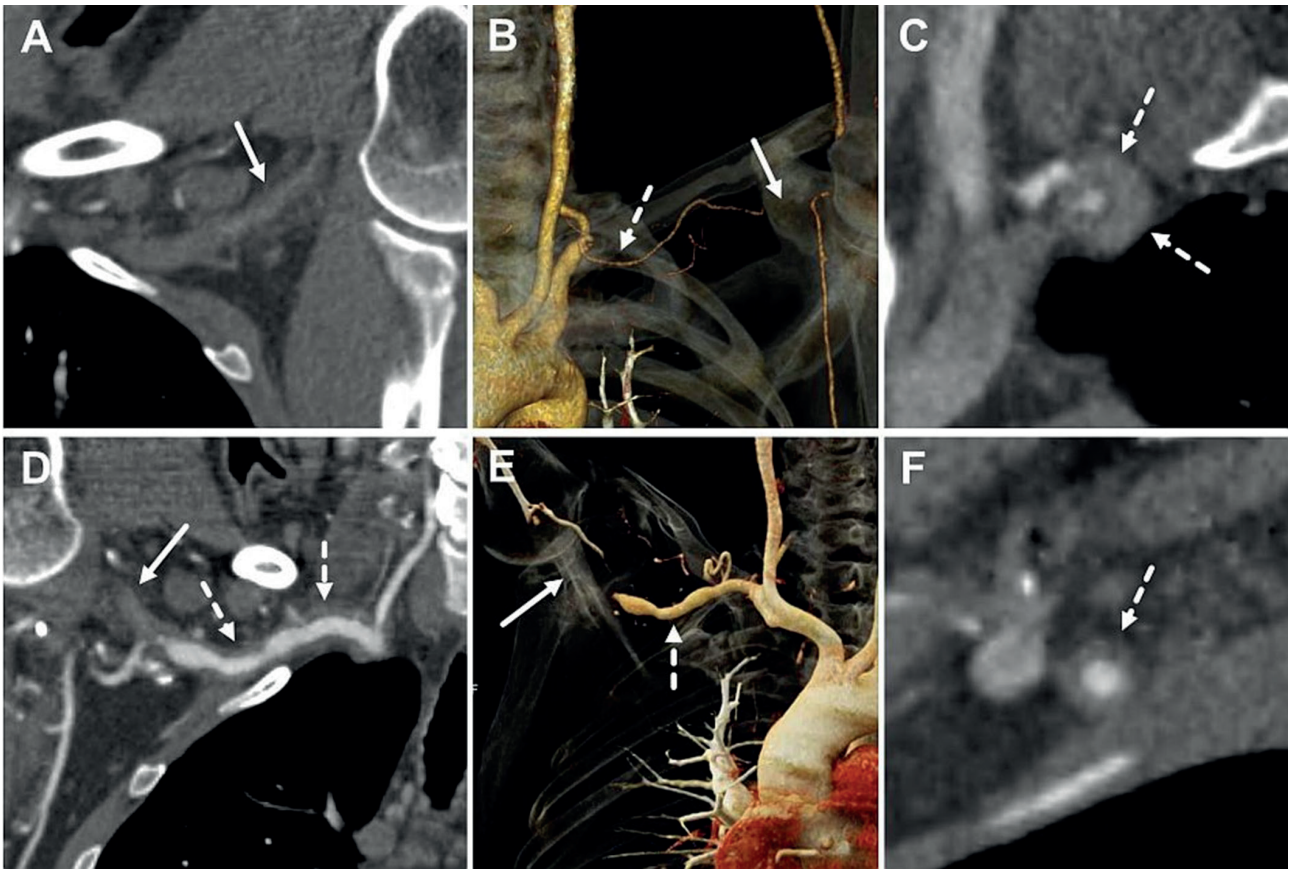

Figure 1. CT angiography images of a 65-year-old male patient with left upper extremity claudication. Coronal MPR (multiplanar reformat) image shows left axillary artery occlusion (arrow) (A), which is better delineated in the coronal VRT (volume-rendered technique) image (arrow)(B). Also, long segment severe stenosis of the left subclavian artery is seen (dashed arrow)(B). Circumferential wall thickening with a "double-ring" sign causing severe stenosis in the left subclavian artery is shown (dashed arrows)(C). Coronal MPR image (D) and cinematic VRT image (E) demonstrate wall irregularities, focal ectatic areas, and concentric wall thickening of the right subclavian artery (dashed arrows). Also, note that the right axillary artery is occluded (arrow). Circumferential wall thickening of the right subclavian artery is observed (dashed arrow)(F).

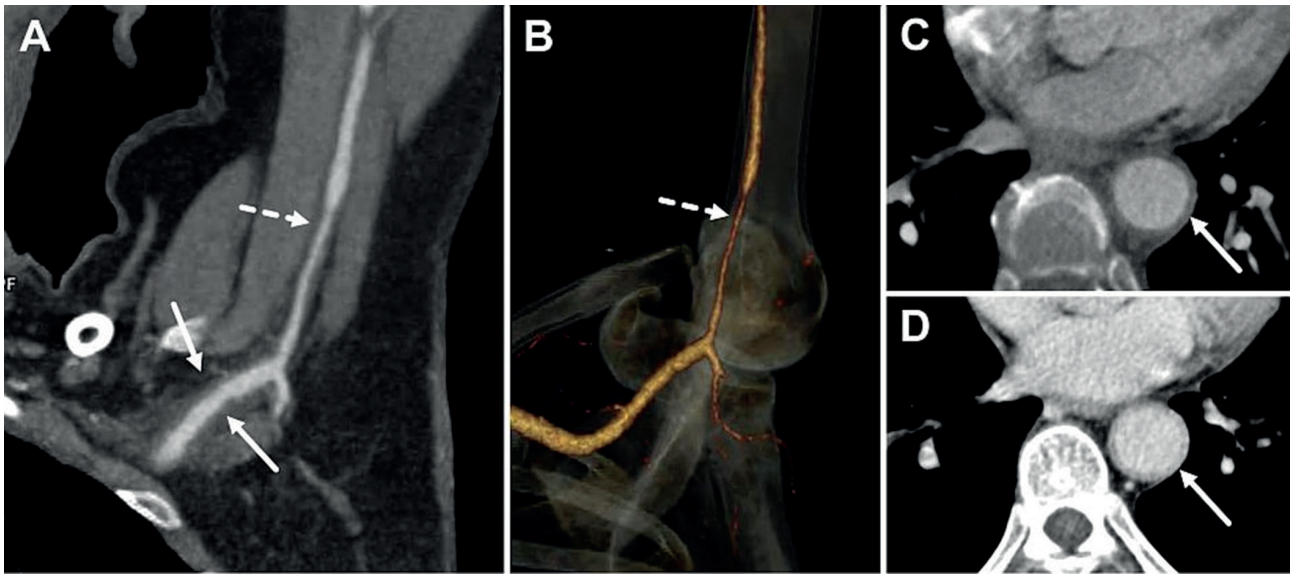

Figure 2. CT angiography of a 67-year-old female patient with temporal arteritis. Coronal MPR (A) image shows concentric wall thickening of left axillary artery (arrows). Also, concentric wall thickening causing severe stenosis in the proximal brachial artery is seen in coronal MPR (A) and VRT (B) images (dashed arrow). Axial arterial phase CT image demonstrates wall thickening of the thoracic aorta (arrow, C) and wall enhancement in venous phase image (arrow, D).

Aortic dilatation- mainly in the thoracic aorta- is reported in nearly 15% of GCA patients at the time of diagnosis [7].

Lariviere et al. compared CTA and FGD-PET/CT for the diagnosis of GCA and reported sensitivity rates of 73% and 66.7%, respectively. Although sensitivity rates were similar, CTA had a lower specificity (84.6%) compared to the uptake on FDG-PET/CT (100%) [17]. In De Boysson et al.'s study, CTA has a sensitivity of 95% and a specificity of 100% in a per-patient analysis when FDG-PET/CT is accepted as a reference [18]. In both studies, FDG-PET/CT had a higher performance in per-segment analysis, especially for the aorta branches [17,18].

MR angiography in LV-GCA

MR angiography (MRA) is increasingly preferred in diagnosing and following large-vessel vasculitis due to the lack of ionizing radiation. MRA should be obtained covering the entire area between carotid bifurcations and iliac arteries, including axillary and brachial arteries [3]. Multiplanar dark blood (HASTE) and bright blood (single shot true-FISP) images can be acquired for the morphological assessment of vessel walls. Also, short-tau inversion-recovery (STIR) and T2 weighted fast spin-echo images allow the identification of vessel wall edema without using contrast material. After administering gadolinium-based contrast agents, three-dimensional (3D), MR angiography for the detection of luminal changes and post-contrast T1 weighted imaging for the assessment of wall enhancement should be performed [13,14,19].

MRA has a reported sensitivity of 79% and specificity of 96% for the GCA diagnosis [20]. Similar to CTA, imaging findings of LV-GCA are concentric wall thickening, wall edema, wall enhancement, and luminal changes such as stenosis, occlusion, and aneurysms [21,22](Figure 3). Although the wall edema on T2-weighted sequences has been attributed to disease activity, there are controversial observations in the literature stating the persistency of edema despite clinical remission [22–24]. Also, EULAR said that T2-weighted imaging for edema is more prone to artifacts and, therefore, less sensitive [3].

MRA has disadvantages such as longer acquisition time and lower spatial resolution compared to CTA. Also, patient cooperation, examination protocol, and operator dependence affect the examination's quality [25].

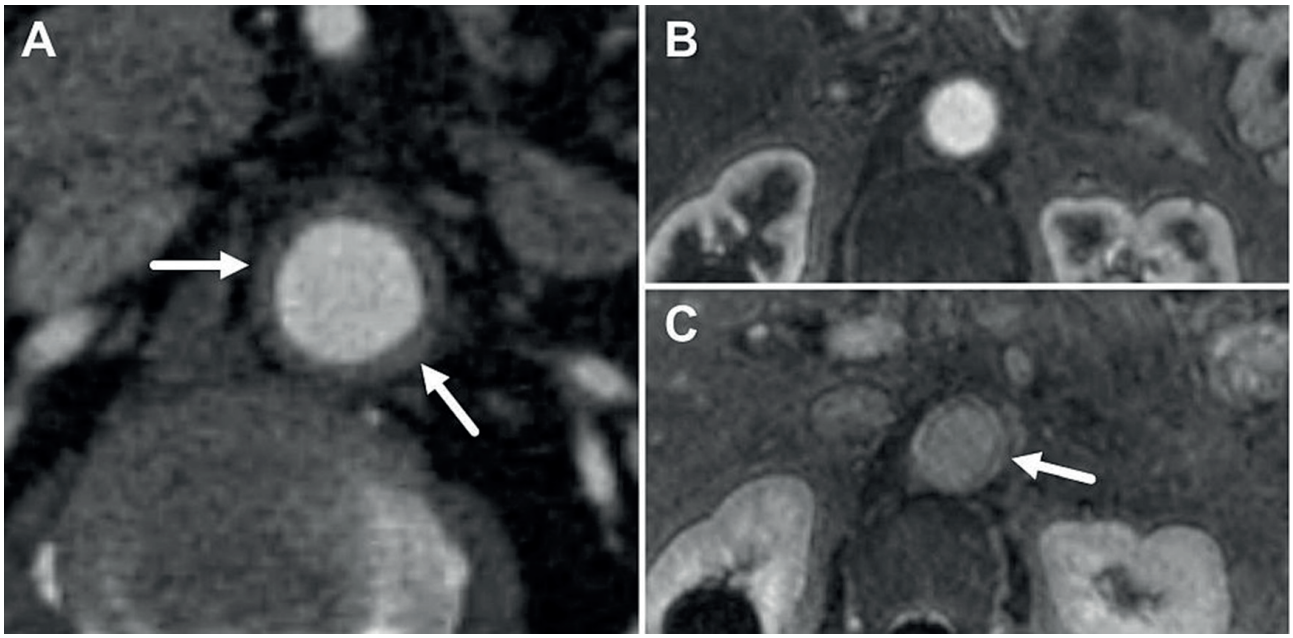

Figure 3. CT and MR angiography images of the same patient as in Figure 1. Axial CT image in the arterial phase shows circumferential wall thickening of the abdominal aorta (arrows). The thickened abdominal aorta wall demonstrates enhancement in the delayed phase MRA image (arrow, C), which is better depicted when compared to the arterial phase image (B).

Differentiation of LV-GCA from atherosclerosis based on imaging

Since GCA typically affects the elderly, atherosclerosis is an important differential diagnosis that should be considered in this patient group. Moreover, atherosclerosis may accompany LV-GCA, and imaging findings may be confusing.

Vessel wall changes are also observed in atherosclerosis, mainly affecting intima. In contrast to circumferential and rather long segment wall thickening of GCA, atherosclerosis mainly presents with focal/patchy, eccentric wall thickening resulting in plaque formations [8,11]. Vessel wall calcifications on CTA are commonly associated with atherosclerosis. Still, calcifications do not rule out vasculitis as they may also occur in large-vessel vasculitis in the long-term [26] (Figure 4).

The affected vascular structures may aid in differentiating atherosclerosis from LV-GCA. LV-GCA most commonly affects the supra-aortic branches of the aorta, such as the axillary - subclavian artery and thoracic segment of the aorta. In contrast, atherosclerosis favors carotid arteries at the supra-aortic level, abdominal aorta, and iliofemoral arteries [27]. Contrast enhancement in the vessel wall is generally not expected in atherosclerotic lesions. However, as atherosclerosis is also an inflammatory condition, contrast enhancement, and FDG uptake may be observed in inflamed plaques [11,28,29].

Follow-up imaging in LV-GCA

According to EULAR recommendations, imaging can monitor long-term results and complications of GCA, such as vessel occlusions, stenoses, or aneurysm formation. There is no clear recommendation regarding which modality should be used in the follow-up process. As the modality selection depends on the availability and local expertise, the choice of imaging frequency and modality in follow-up should be decided on a patient basis. When a flare is suspected, imaging findings may help confirm disease activity. On the other hand, imaging is not recommended in the case of clinical and biochemical remission [3]. Although the wall enhancement mostly disappears after treatment, this may occur later than clinical

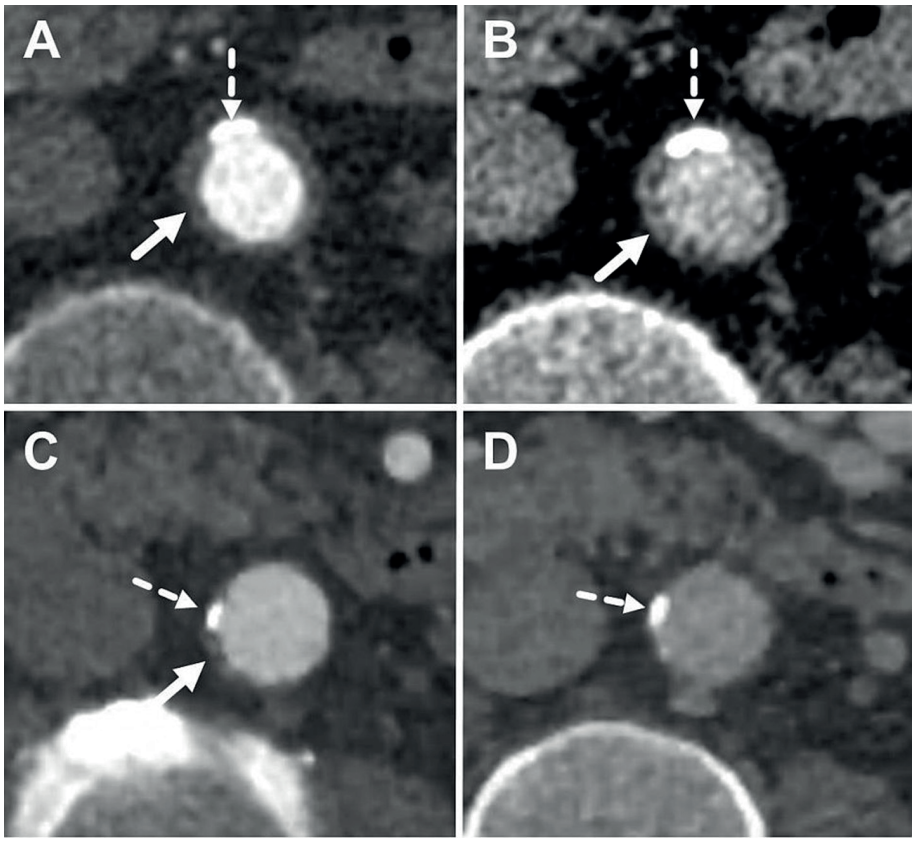

Figure 4. Differentiation of GCA from atherosclerosis. Axial arterial (A) and venous phase (B) CT images of a patient with GCA show concentric wall thickening of the abdominal aorta with contrast enhancement in the venous phase (arrows). Calcifications may also occur with large vessel vasculitis (dashed arrows, A-B). Axial arterial (C) and venous phase (D) CT images of a patient with atherosclerosis show eccentric wall thickening without enhancement in atherosclerosis, contrary to vasculitis (arrows). Plaque calcification is also seen (dashed arrows, C-D).

and biochemical remission. Also, even though the number of affected segments and wall thickness decreases, wall thickening may persist in two-thirds of LV-GCA patients [14,30].

Imaging in the follow-up period is vital in assessing potential complications of structural vessel damage. Increased incidence of aortic aneurysms predominantly in the thoracic aorta and aortic dissections were reported in GCA patients in the literature [31]. In Garcia- Martinez et al.'s study, aortic structural damage such as dilatation and aneurysm more commonly in thoracic aorta is observed in 22% of the patients after a median of 5.4 years regardless of clinical activity [32]. These patients were further monitored over a median follow-up for 10.3 years, and significant increases in aortic diameters were observed [33].

Conclusion

The role of CT and MR angiography in the diagnosis of extracranial large-vessel type GCA has gradually increased. By demonstrating the luminal and vessel wall changes of the aorta and aortic branches, imaging may provide important information to support the diagnosis in the presence of clinical suspicion and reveal possible complications in the follow-up. Although the imaging findings may overlap with atherosclerotic changes, type of wall thickening (concentric or eccentric), presence of calcifications or wall enhancement, and distribution of the affected vessels may aid in the differential diagnosis.

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