REVIEW ARTICLE

Imaging findings in Giant Cell Arteritis: Don't Turn A Blind Eye To The Obvious!

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ABSTRACT

Giant cell arteritis (GCA) is the most common primary large vessel systemic vasculitis in the western world. Even though the involvement of scalp and intracranial vessels has received much attention in the neuroradiology literature, GCA, being a systemic vasculitis can involve multiple other larger vessels including aorta and its major head and neck branches. Herein, the authors present a pictorial review of the various cranial, extracranial and orbital manifestations of GCA. An increased awareness of this entity may help with timely and accurate diagnosis, helping expedite therapy and preventing serious complications.

ABBREVIATIONS: ACR= American College of Rheumatology, AION= Anterior Ischemic Optic Neuropathy, EULAR= European League Against Rheumatism, GCA= Giant Cell Arteritis, LV-GCA= Large vessel GCA, PMR= Polymyalgia Rheumatica, US= Ultrasound, VWI= Vessel Wall Imaging.

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1. Introduction

Giant cell arteritis (GCA), categorized as a large vessel vasculitis under the 2012 Revised Chapel Hill Consensus Conference, is the most common primary systemic vasculitis in the western world in people older than 50 years.[1-5] The peak incidence of GCA occurs in individuals in their eighth decade of life. GCA is more common in Scandinavians and North Americans of Scandinavian descent, and more common in women (F:M=2:1).[4, 6, 7] The lifetime risk of developing GCA is about 1% for women and 0.5% for men, with an overall GCA incidence of 20 per 100,000 population in people older than 50 years.[4, 8] Patients may present with a wide range of symptoms, including headache (75%), jaw claudication (30%), swelling or tenderness along the temporal artery (50%), visual (15%) or neurological (30%) symptoms.[4, 5] There is substantial overlap with polymyalgia rheumatica (PMR) and about 40-60% of GCA patients have PMR while about 15-20% of PMR patients have GCA.[8] The precise pathophysiology of GCA is not well defined. Seasonal variations in GCA onset may reflect a role of environmental factors in genetically prone individuals. A genome-wide association study in 2017 showed a strong human leucocyte antigen class II association, besides identifying risk polymorphisms in genes encoding plasminogen and an isoform of the alpha subunit of collagen prolyl 4-hydroxylase, which are consistent with alterations in vascular remodeling in disease susceptibility.[7, 9]

GCA has two broad, overlapping phenotypes. Patients with predominantly cranial GCA (also referred to as C-GCA) often show involvement of branches of external carotid artery, such as superficial temporal, facial or occipital artery etc. A recent ultrasound-based study noted that superficial temporal artery involvement was more common (76%), followed by facial (41%) and occipital (31%) arteries. On the other hand, patients with large vessel GCA (LV-GCA) are more likely to have aortic and upper extremity arterial involvement, with axillary arteries being most frequently involved.[7, 10] GCA may classified based on the previously outlined criteria by the American college of Rheumatology (ACR) in 1990 which largely relied on clinical, lab and pathological abnormalities for GCA diagnosis. However, these have been criticized for their poor sensitivity and exclusion of extra-cranial large vessel involvement.[3, 5, 11] For example, Muratore et al, in their study, noted that while 95% of patients with cranial GCA met at least three ACR criteria needed for diagnosis, only 39% of patients with LV-GCA satisfied at least three criteria.[11] Similarly, temporal artery biopsy (TAB) which is considered the 'gold standard' for diagnosis has low sensitivity.[5] Moreover, the widespread use of non-invasive vascular imaging has further necessitated need for revised criteria to reflect current practice.

The 2022 ACR/ European League Against Rheumatism (EULAR) updated GCA classification criteria to include: positive temporal artery biopsy (TAB) or temporal artery halo sign on ultrasound (+5); erythrocyte sedimentation rate (ESR) ≥50 mm/hour or C reactive protein ≥10 mg/L (+3); sudden visual loss (+3); morning stiffness in shoulders or neck, jaw or tongue claudication, new temporal headache, scalp tenderness, temporal artery abnormality on examination, bilateral axillary involvement on imaging and fluorodeoxyglucose–positron emission tomography activity throughout the aorta (+2 each). A cumulative score of ≥6 points was shown to achieve a sensitivity of 87.0% (95% CI 82.0% to 91.0%) and specificity of 94.8% (AUC: 0.91; 95% CI 0.88 to 0.94) for GCA diagnosis in the validation cohort (table-

1).[12] It is important to note that application of these criteria should only be considered once a diagnosis of vasculitis has been made and alternate diagnoses have been excluded.

Before the introduction of corticosteroid therapy, the estimated mortality rate among GCA patients was approximately 12.5%. However, with appropriate treatment, the long-term outcomes and survival rates are similar to age matched population.[4] Important complications in GCA include vision loss (up to 15%), aortic aneurysms (10-15%) and dissections, and cerebrovascular events (2-4%).[8, 11, 13]

Typical histopathological findings in GCA include vessel wall inflammation, intimal thickening, and internal elastic lamina fragmentation (**Fig 1**). Multinucleated giant cells are seen in only about half of the cases. Other findings include lympho-mononuclear predominant panarteritis and inflammation of the vasa vasorum.[4, 8] Presence of fibrinoid necrosis often implies alternate diagnosis such as ANCA associated vasculitis.[8]

No single imaging modality is generally sufficient to evaluate disease extent and severity in GCA. For the same reason, the 2023 EULAR update on imaging recommendations suggest different imaging modalities for cranial and extra-cranial GCA. The guidelines also make suggestions for technical and operational parameters for the various imaging modalities which may be useful for designing and implementing imaging protocols at institutional level.[14] Besides the recommendations, the paper also outlines three overarching principles, namely a) performing early imaging in suspected GCA which should not impede treatment initiation, b) imaging by trained experts using standardized protocols and c) avoiding additional testing in patients with high clinical suspicion and positive initial imaging as well as patients with low clinical suspicion and negative imaging findings.[14]

Even though imaging in GCA has largely focused on the scalp and extra-cranial vessels, additional imaging findings in the orbits, temporalis muscle and intracranial vessels have also been reported.[3, 5] Herein, we review the previously reported imaging findings in GCA, which can be helpful in accurate and timely detection of this systemic large vessel vasculitis.

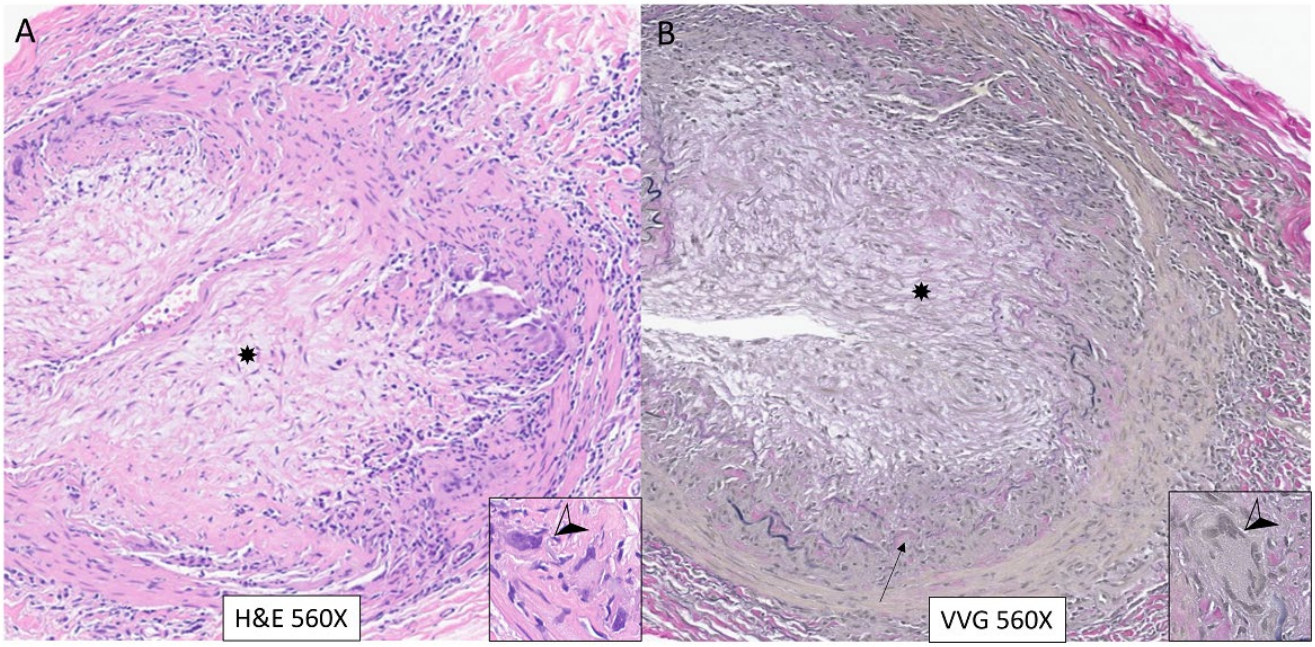


FIG 1. H&E (A) and Verhoeff-van Giesson (VVG) (B) stain photomicrographs from a temporal artery biopsy in a positive GCA case reveals severe arteritis with inflammatory lymphocytic cells throughout the vessel wall (A, B). There is loss of the internal elastic membrane (B, arrow) with marked intimal fibroplasia (Asterix) resulting in complete obliteration of lumen. Inserts with magnified views show multinucleated giant cells (arrowheads) interspersed between lymphocytes.

Table 1: Updated classification criteria for GCA diagnosis.

CLASSIFICATION CRITERIA FOR GIANT CELL ARTERITIS *
2022 American College of Rheumatology (ACR) & European Alliance of Associations for Rheumatology (EULAR)

ABSOLUTE REQUIREMENT

- **Age ≥ 50 years at time of diagnosis**

ADDITIONAL CLINICAL CRITERIA

• **Morning stiffness in shoulder/neck area**	**+2**
• **Sudden visual loss**	**+3**
• **Jaw or tongue claudication**	**+2**
• **New temporal headache**	**+2**
• **Scalp tenderness**	**+2**
• **Abnormal examination of temporal artery**	**+2**

LABORATORY, IMAGING & BIOPSY CRITERIA

• **Maximum ESR ≥ 50 mm/hr or maximum CRP ≥ 10mg/liter**	**+3**
• **Positive temporal artery biopsy or halo sign on ultrasound**	**+5**
• **Bilateral axillary involvement**	**+2**
• **FDG-PET activity throughout aorta**	**+2**

Sum the scores for 10 items, if present. A score of ≥ 6 points is needed for diagnosis of giant cell arteritis

***CONSIDERATION WHILE APPLYING THE CRITERIA**

- **Classification criteria should be applied when a diagnosis of medium-vessel or large-vessel vasculitis has been made**
- **Alternate diagnosis mimicking vasculitis should be excluded prior to applying the criteria**

Adapted from Ponte C et al & DCVAS Study Group. 2022 American College of Rheumatology/EULAR Classification Criteria for Giant Cell Arteritis. Arthritis Rheumatol. 2022 Dec;74(12):1881-1889.

2. Vascular findings in cranial GCA

Most studies have broadly focused on involvement of the superficial temporal artery branches with some studies additionally evaluating intracranial and orbital vessels. As per the most recent EULAR recommendations, ultrasound (US) of the temporal and axillary arteries is recommended as the first-line imaging modality in patients with suspected, predominantly cranial GCA, with a non-compressible 'halo-sign' being the most suggestive finding.[14, 15] Additional imaging findings may include vascular stenosis and/or occlusion. The halo sign refers to the presence of homogenous, hypoechoic wall thickening which is concentric on transverse images (**Fig 2**). The halo sign has a sensitivity of 68% and a specificity of 91% for GCA diagnosis. Its specificity reaches up to 100% when present bilaterally.[3, 4] A recent metanalysis only using studies with low risk of bias noted a pooled sensitivity of 88% (95% CI 82-92%) and specificity of 96% (95% CI 86-99%) for US in GCA diagnosis.[15] The halo sign may also be seen in other involved vessels.[10]

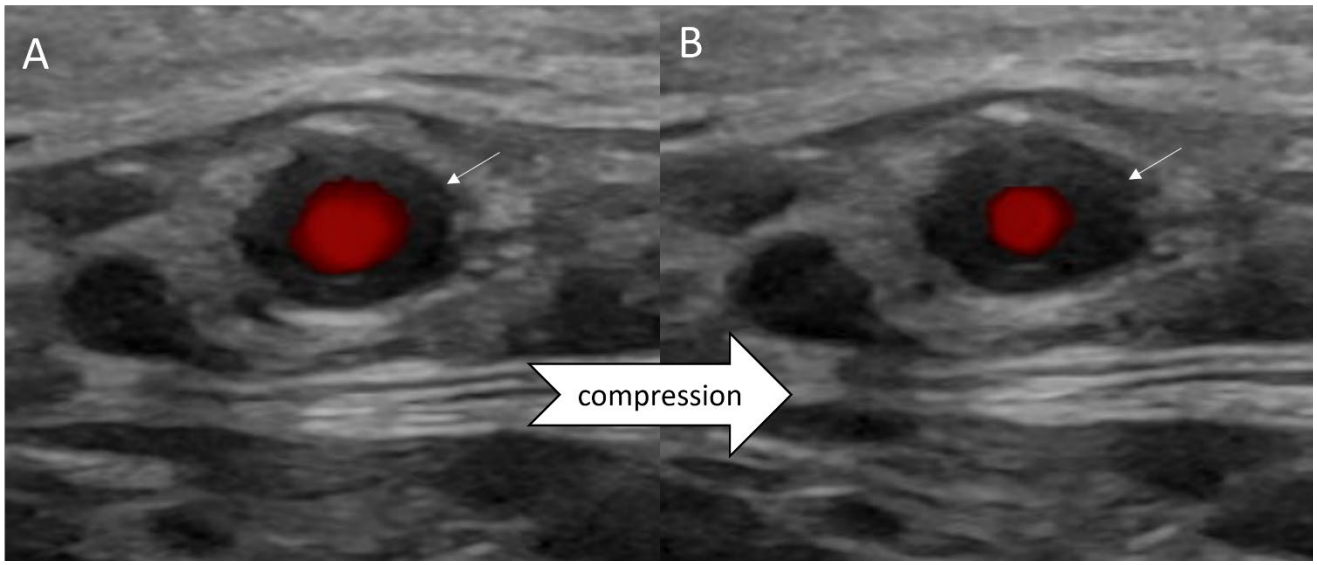


Fig 2: “Halo” sign and compressibility test in a patient with biopsy proven GCA. Ultrasound images (A,B) show a dark hypoechoic halo around the superficial temporal artery (STA) lumen (arrows) representing the vessel wall inflammation with partial loss of normal compressibility and decrease in flow.

US evaluation of the vessels should ideally include the common temporal arteries and their frontal and temporal branches, as well as the axillary arteries. This is preferably performed with linear probes with at least 15-18 MHz and ≥12-15 MHz frequencies for temporal and axillary arteries respectively.[3] Additional evaluation of facial, vertebral, occipital, subclavian and femoral arteries may be helpful when the GCA diagnosis is not clear. Well recognized advantages of US include easier availability, non-invasive nature and lack of any radiation or need for intravenous contrast. The main limitation is operator dependence.[3, 16]

Several recent studies have also evaluated the utility of high-resolution MRI vessel wall imaging (VWI) for cranial GCA, which is generally performed using standard gadolinium dose contrast, a T1-based sequence using fat-suppression and a five-minute delay between contrast injection and image acquisition.[1, 5, 13] The reported imaging findings in GCA cases include thickening and enhancement along the vessel walls of the superficial temporal artery and its branches, as well as the occipital artery. The enhancement is generally concentric and can be segmental (**Fig 3**).[1, 17, 18] The enhancement may be semi-quantitatively assessed based on the previously described four-point scale by Klink et al, with scores of 0-1 representing physiologic features and scores of 2-3 reflecting vessel wall involvement.[1] Siemonsen et, al., noted that most patients with GCA showed clear signs of mural inflammation in at least two affected vascular segments.[18] Luminal stenosis and diffusion restriction along the involved vessel segments may also be seen (**Fig 4**).[3, 19] With treatment, changes of vasculitis improve, though residual wall thinning and pseudoaneurysm may be seen (**Fig 5**). The reported sensitivity and specificity of MRI-VWI on the more recent metanalyses was 91% and 78% respectively, when compared with temporal artery biopsy as the reference standard.[5] MR-VWI does have a high negative predictive value for cranial GCA and a normal MR-VWI study may imply a low probability of a positive TAB.[20] The 2023 update to the EULAR guidelines recommends both high resolution MRI and FDG-PET as alternatives to US for assessment of cranial arteries in suspected GCA.[14]

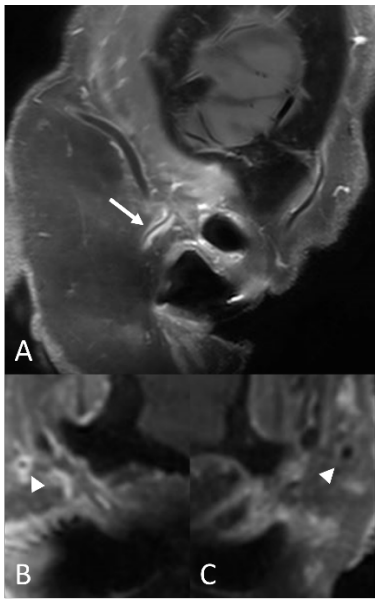


Fig 3: Sagittal post contrast T1-SPACE image shows circumferential wall enhancement along the right superficial temporal artery (STA,

arrow, A). Oblique MPR images perpendicular to the involved vessel (in A) show corresponding circumferential enhancement (arrowhead, B). The left STA (arrowhead, C) is normal.

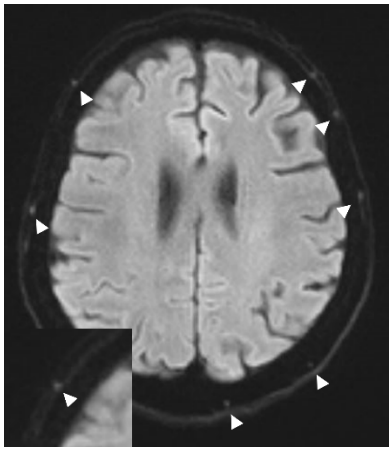


Fig 4: Axial DWI images in a treatment naïve GCA patient showing scattered foci of increased diffusion signal along bilateral scalp vessels (arrowheads). Inset in bottom left shows magnified DW signal changes along the right frontal STA branch.

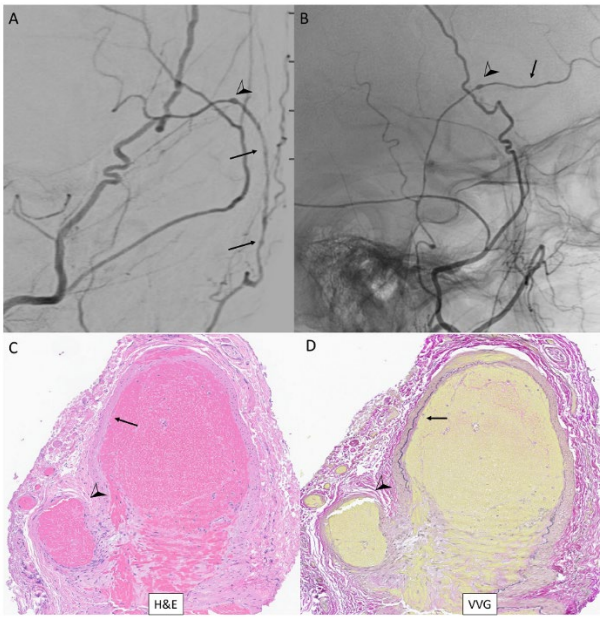


Fig 5: Long-standing GCA in a 66-year-old woman with pseudoaneurysm of the superficial temporal artery. Frontal (A) and lateral (B) projections on catheter angiogram (ECA injection) reveal multifocal areas of narrowing involving the frontal and parietal branches of STA (A, B, arrows). Small pseudoaneurysm is noted along the frontal branch of STA (A, B, arrowheads). Temporal artery biopsy with H&E (C) and VVG (D) stains reveals marked atrophy of the arterial wall (C, D arrows) suggesting healing with pseudoaneurysm (C, D, arrowheads).

In general, MRI-VWI evaluation at higher magnet strength is more fruitful and 3D-VWI sequences perform better in detecting GCA changes, being more specific when compared to 2D-VWI sequences (91% vs 84%), with similar overall sensitivity (70 vs 72%).[5, 17] 3D-VWI sequences also have additional advantages of larger field-of-view, ability to generate reformatted images without loss of image resolution and evaluation of multiple vessels along their course.[17]

A cross-sectional study recently compared US and MRI for GCA in patients with both newly diagnosed and established disease. The authors noted that in this small patient cohort, US detected vasculitic changes more frequently than MRI (37% vs 21%, $p < 0.001$) and was also more sensitive in detecting vasculitic changes in larger head and neck vessels. However, the lack of vasculitic changes on MRI/MRA was significantly associated with disease remission. With US, vasculitic changes were noted in both active and inactive disease. Notably, the study used 1.5T MRI and larger head and neck vessels (such as carotid, axillary and subclavian vessels) were only assessed on MRA, and not MRI.[16]

When compared to US, the main disadvantages of MR-VWI include lack of wider availability, requirement of intravenous contrast, need for subspecialty expertise in image evaluation and generally longer wait times. The latter is especially important as the imaging findings can quickly improve after high-dose corticosteroid administration, thus reducing diagnostic sensitivity.[1, 3, 4] MR-VWI is also more prone to artifacts from venous or slow flow and non-suppression of intraluminal signal which can be occasionally problematic (**Fig 6**). Finally, MR imaging may be contraindicated due to patient specific factors (such as cochlear implants, aneurysm clips and so forth).[16]

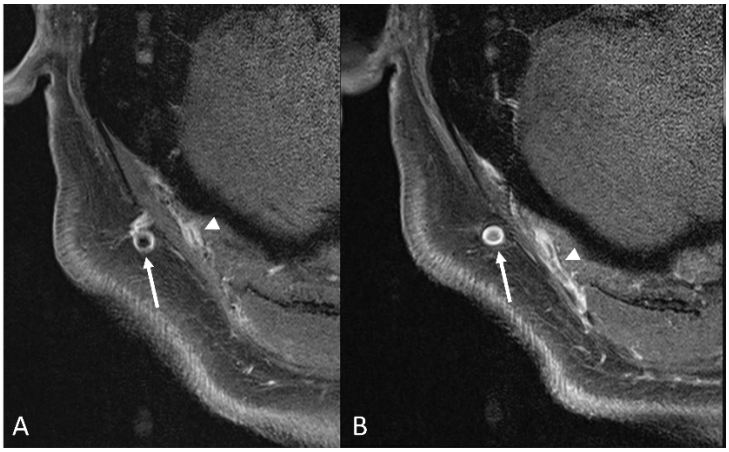


Fig 6: Axial post-contrast images reveal flow artifacts in the right occipital vein (arrows), which should not be misinterpreted as mural inflammatory changes. There is circumferential enhancement along the right superficial occipital artery (arrowheads).

However, MR-VWI has additional advantages of allowing simultaneous evaluation of orbits, temporalis muscles and intracranial vessels.[5] The latter may be affected in about 10-15% (reported up to 50% in some studies) of GCA patients, with intradural ICA and vertebral vessels most commonly involved. The involved vessels may show circumferential wall thickening and enhancement, similar to extra-cranial counterparts.[13, 18, 21] A recent study noted that most lesions involve greater than 5 mm of vessel length with none showing >70% luminal stenosis.[13]

There is limited literature on role of cranial CT angiography in GCA. Conway et. al., previously retrospectively evaluated CTA head studies in a cohort of fourteen treatment naïve patients who were subsequently diagnosed with GCA, along with similar number of age-matched controls. Blurred vessel margins and perivascular enhancement was noted in ten cases and two controls, yielding an accuracy of about 78.6% for CTA (**Fig 7**). Interestingly, the presence of STA occlusion, stenosis or calcification was not significantly different between the two groups.[22]

More recently, studies using Fluorine-18-fluorodeoxyglucose positron emission tomography (FDG-PET) have also shown promising results in cranial GCA diagnosis (**Fig 8**). Nielsen et. al., studied a cohort of 44 patients with equal number of age-matched controls. Based on presence or absence of FDG uptake in the temporal, maxillary and vertebral arteries, PET-CT had 82% sensitivity and 100% specificity.[23] Limitations of FDG-PET include restricted availability, high cost and radiation.[3]

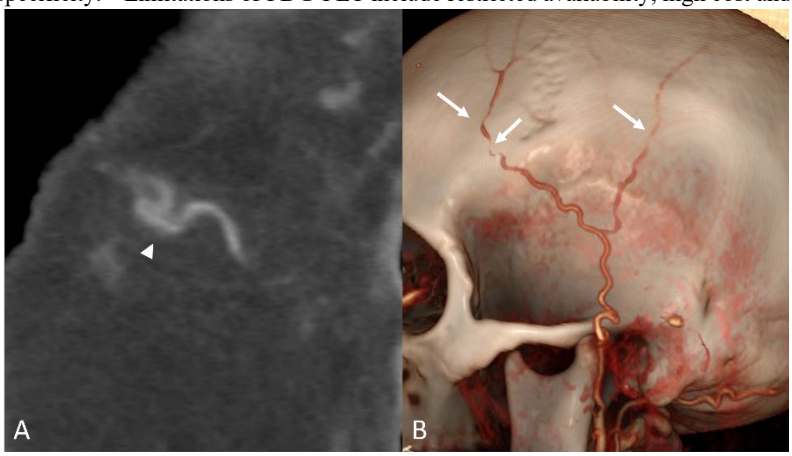


Fig 7: Sagittal CTA-MPR image (A) reveals blurring of vessel margins and subtle fat stranding along the frontal branch of the right STA. Volume rendered image (B) shows scattered areas of vessel irregularity and stenosis along the STA branches (arrows).

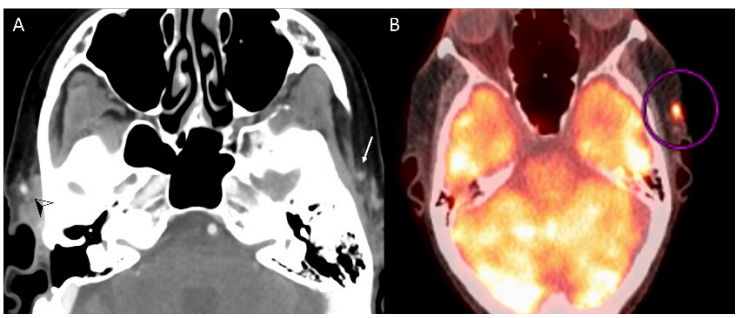


Fig 8: Axial CTA image (A) in a patient with newly diagnosed GCA shows reduced contrast opacification along the left STA (arrow), and a normal appearing right STA (arrowhead). Fused PET-CT image (B) from the same patient at a slightly cranial level shows prominent radiotracer uptake on the left (circled).

3. Vascular findings in large vessel GCA

In comparison to cranial GCA, LV-GCA predominantly involves the thoracic aorta and aortic arch branches, with or without cranial vessel involvement.[3, 24] Kermani et. al., in their prospective, longitudinal, multi-center study noted that 66% of GCA patients had at least one LV arterial lesion at diagnosis and 39% of those with follow-up imaging developed new lesions, often in the first two years. All patients with new lesions had baseline imaging abnormalities.[25] LV-GCA patients tend to behave slightly differently than patients with cranial GCA, and are younger at presentation, have longer symptom duration prior to diagnosis, are more likely to have associated polymyalgia rheumatica, higher incidence of relapses and require longer corticosteroid treatment. These patients often have fewer cranial symptoms, vision loss and are generally more TAB negative.[11, 24]

Frequently involved vessels include aorta, subclavian, axillary and brachial arteries, with involvement of lower extremity arteries being less common.[11] The 2023 update to the EULAR guidelines recommends use of FDG-PET as the preferred modality for evaluation of extracranial arteries, with a sensitivity of 76% and specificity of 95% with the clinical criteria as reference standard. There is overall limited evidence on the utility of CTA and MRA in LV-GCA.[14]

On FDG-PET, areas of active vasculitis show increased radiotracer uptake along the vessel wall/ course (**suppl fig 1**). Besides evaluating the presence or absence of inflammation, FDG-PET is also helpful to determine overall extent of vasculitis and simultaneously exclude presence of underlying malignancy and infection. Like MRI, the diagnostic accuracy can drop considerably in treated patients; therefore, imaging very early in the disease course is essential.[3, 4, 14]

CTA and MRA can also be used to evaluate LV-GCA, and demonstrate wall thickening, enhancement and long segment tapering stenosis along upper extremity vessels, the latter being present in 3-15 % of cases (**suppl fig 2**).[4, 8] Underlying aortitis most commonly involves the thoracic aorta and may be clinically silent.[4] Espitia et. al., noted aortic complications in 23.5% of their LV-GCA patients, predominantly consisting of aneurysms and dissections. These were seen after a median delay of 27 months post-diagnosis and were significantly more common in patients with symptomatic aortitis, defined as presence of dorsal/ lumbar/ abdominal pain and/or aortic insufficiency.[26] Quinn et. al., compared FDG-PET and MRA in LV vasculitis (including GCA and Takayasu arteritis) and noted that MRA outperformed FDG-PET for evaluating disease extent, but had lower inter-reader correlation. Even though clinical status was more closely correlated with FDG-PET activity, about 51% of patients with LV vasculitis in clinical remission had active disease by both MRA and FDG-PET.[27]

4. Orbital findings in GCA

Vision loss remains one of the most dreaded complications of GCA, often occurring suddenly and painlessly. It may be unilateral or bilateral, with a higher risk of bilateral involvement if the unilateral vision loss is not emergently treated with high-dose corticosteroids.[4, 8]

Inflammatory involvement of intra-orbital structures have been reported in about a third of GCA patients, most commonly along the optic nerve sheaths followed by the ophthalmic artery and intraconal fat (**Fig 9**).[28] Gospe et al, also noted that both intracranial ICA and optic nerve sheath enhancement were observed in patients with TAB+ GCA and a combination of these two imaging findings was highly specific for GCA.[29] Similarly, Sommer et. al., noted that MRI-VWI showed bilateral orbital involvement in 50% of cases with arteritic anterior ischemic optic neuropathy (AION) when only unilateral corresponding changes were noted at fundoscopy, suggesting improved detection of subclinical disease and patients at risk of further vision loss.[30] Another study noted that inflammatory changes along the ophthalmic artery were present in all cases with arteritic AION, but in none with non-arteritic AION.[31] Finally, Remond et al, noted that all patients with GCA-AION showed optic nerve head enhancement (central bright spot sign).[32] Similar findings were also noted in about 50% of patients with non-arteritic AION, while none of the healthy controls showed optic nerve head enhancement. The authors postulated that absence of central bright spot sign may suggest underlying non-arteritic AION.

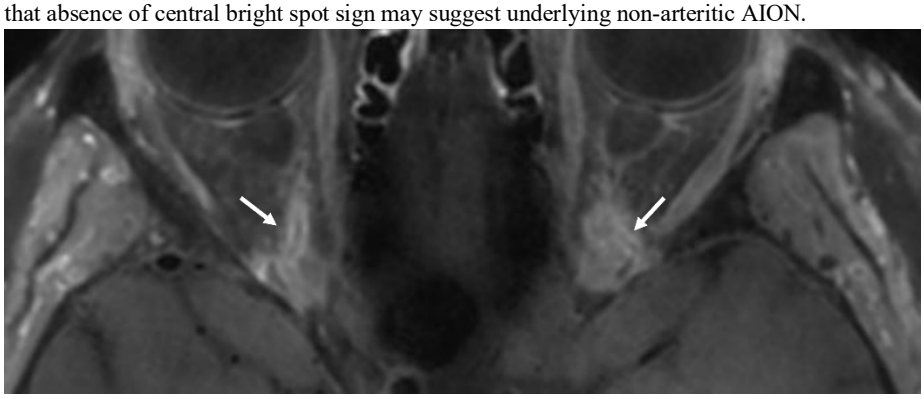


Fig 9: Axial T1-SPACE post contrast image through the orbits in a patient with newly diagnosed GCA shows bilateral retrobulbar intraconal enhancement near the apex with involvement of bilateral ophthalmic arteries (arrows).

5. Miscellaneous findings in GCA

Additional findings in GCA patients, as described on MRI, include temporalis muscle inflammation (about 20%), and vasculitis of the deep temporal artery (34-49%), with simultaneous involvement of both structures reported in about 20% of patients (**Fig 10**). The latter shows moderate correlation with jaw claudication.[6]

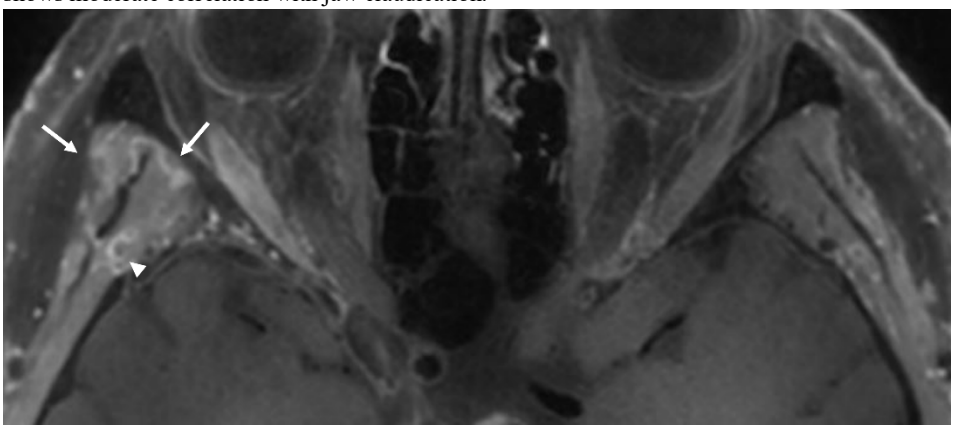


Fig 10: Axial T1-SPACE post contrast image (same patient as fig 9) shows asymmetrically increased enhancement along the right temporalis muscle (arrows) and along the deep temporal artery (arrowhead).

6. Challenges and Future Directions:

A recent population-based cohort study noted that even though the incidence of GCA remained constant over the past two decades (1996-2018), the proportion of GCA patients receiving TAB declined sharply from 70-80% to 29-39% after 2016, while the use of diagnostic imaging increased from 2% to 66% between 2000-2018, underscoring the role of non-invasive imaging in GCA diagnosis.[33]

Despite the increasing role of imaging in GCA diagnosis, its utility in follow-up remains a topic of intense research. Koster et. al., for example noted that there was a discordance between imaging findings and clinical symptoms, especially during follow-up.[24] Another study noted that even though Tocilizumab led to complete clinical and biochemical remission in their cohort, imaging abnormalities of the extracranial large arteries only normalized in a third of the patients.[34] On the other hand, treatment rapidly improves superficial cranial vessels and mural inflammatory changes such as intima-media thickness, contrast enhancement and mural thickening.[1, 35] For these reasons, the added value of imaging in response assessment, to define remission, in predicting short and long-term outcomes and its association with novel laboratory markers remains under investigation.[14]Similarly, the use of imaging findings as an outcome tool needs to be prospectively evaluated in randomized controlled trials.

Additionally, some recent studies have shown that concurrent evaluation of cranial and LV-GCA improves overall sensitivity without negatively impacting specificity of GCA diagnosis with both US and PET-CT. The diagnostic accuracy of combined cranial and LV-GCA with MRI remains under investigation.[36] Finally, some recent studies have shown promising results in terms of diagnosis and management of GCA, using artificial intelligence based methods, with either imaging or non-imaging (patient) data.[37, 38] These however need to be prospectively validated on larger patient cohorts.

In conclusion, GCA can have a spectrum of imaging manifestations involving both cranial and extra-cranial sites. Imaging plays an increasingly important role in timely and accurate diagnosis. A broader recognition of its imaging abnormalities and awareness of its protean manifestations may help with prompt initiation of therapy and avoid serious complications.

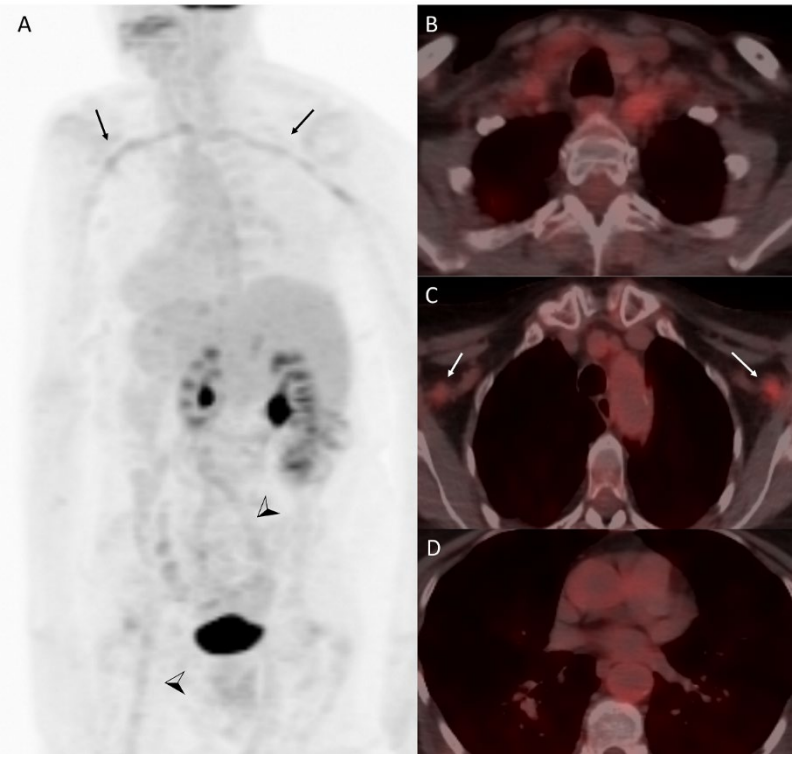
REFERENCES

1. Klink T, Geiger J, Both M, et al. Giant cell arteritis: diagnostic accuracy of MR imaging of superficial cranial arteries in initial diagnosis-results from a multicenter trial. *Radiology* 2014;273:844-852
2. Jennette JC, Falk RJ, Bacon PA, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013;65:1-11
3. Prieto-Pena D, Castaneda S, Martinez-Rodriguez I, et al. Imaging Tests in the Early Diagnosis of Giant Cell Arteritis. *J Clin Med* 2021;10
4. Younger DS. Giant Cell Arteritis. *Neurol Clin* 2019;37:335-344
5. Zhang KJ, Li MX, Zhang P, et al. Validity of high resolution magnetic resonance imaging in detecting giant cell arteritis: a meta-analysis. *European radiology* 2022;32:3541-3552
6. Veldhoen S, Klink T, Geiger J, et al. MRI displays involvement of the temporalis muscle and the deep temporal artery in patients with giant cell arteritis. *European radiology* 2014;24:2971-2979
7. Pugh D, Karabayas M, Basu N, et al. Large-vessel vasculitis. *Nat Rev Dis Primers* 2022;7:93
8. Ameer MA, Peterfy RJ, Khazaeni B. Giant Cell Arteritis (Temporal Arteritis). *StatPearls*. Treasure Island (FL); 2024

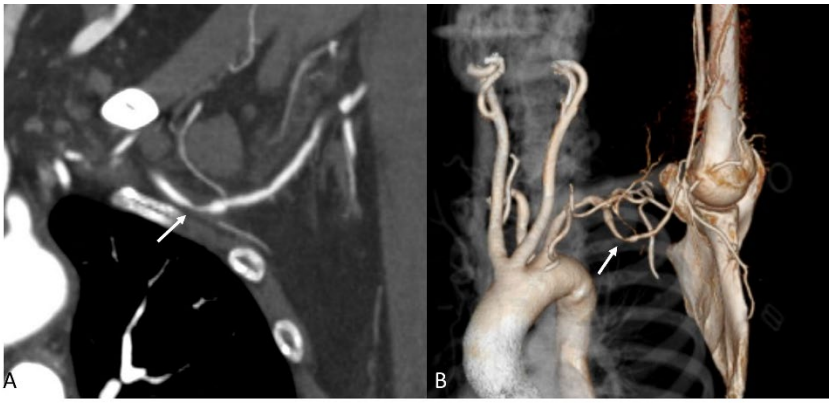
9. Carmona FD, Vaglio A, Mackie SL, et al. A Genome-wide Association Study Identifies Risk Alleles in Plasminogen and P4HA2 Associated with Giant Cell Arteritis. *Am J Hum Genet* 2017;100:64-74
10. Jese R, Rotar Z, Tomsic M, et al. The role of colour doppler ultrasonography of facial and occipital arteries in patients with giant cell arteritis: A prospective study. *European journal of radiology* 2017;95:9-12
11. Muratore F, Kermani TA, Crowson CS, et al. Large-vessel giant cell arteritis: a cohort study. *Rheumatology (Oxford)* 2015;54:463-470
12. Ponte C, Grayson PC, Robson JC, et al. 2022 American College of Rheumatology/EULAR Classification Criteria for Giant Cell Arteritis. *Arthritis Rheumatol* 2022;74:1881-1889
13. Guggenberger KV, Vogt ML, Song JW, et al. High-resolution Magnetic Resonance Imaging visualizes intracranial large artery involvement in Giant Cell Arteritis. *Rheumatology (Oxford)* 2024
14. Dejaco C, Ramiro S, Bond M, et al. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice: 2023 update. *Ann Rheum Dis* 2023
15. Bosch P, Bond M, Dejaco C, et al. Imaging in diagnosis, monitoring and outcome prediction of large vessel vasculitis: a systematic literature review and meta-analysis informing the 2023 update of the EULAR recommendations. *RMD Open* 2023;9
16. Yip A, Jernberg ET, Bardi M, et al. Magnetic resonance imaging compared to ultrasonography in giant cell arteritis: a cross-sectional study. *Arthritis Res Ther* 2020;22:247
17. Poillon G, Collin A, Benhamou Y, et al. Increased diagnostic accuracy of giant cell arteritis using three-dimensional fat-saturated contrast-enhanced vessel-wall magnetic resonance imaging at 3 T. *European radiology* 2020;30:1866-1875
18. Siemonsen S, Brekenfeld C, Holst B, et al. 3T MRI reveals extra- and intracranial involvement in giant cell arteritis. *AJNR Am J Neuroradiol* 2015;36:91-97
19. Seitz L, Bucher S, Butikofer L, et al. Diffusion-weighted magnetic resonance imaging for the diagnosis of giant cell arteritis - a comparison with T1-weighted black-blood imaging. *Rheumatology (Oxford)* 2023
20. Rheaume M, Rebello R, Pagnoux C, et al. High-Resolution Magnetic Resonance Imaging of Scalp Arteries for the Diagnosis of Giant Cell Arteritis: Results of a Prospective Cohort Study. *Arthritis Rheumatol* 2017;69:161-168
21. Donaldson L, Nanji K, Rebello R, et al. Involvement of the intracranial circulation in giant cell arteritis. *Can J Ophthalmol* 2020;55:391-400
22. Conway R, Smyth AE, Kavanagh RG, et al. Diagnostic Utility of Computed Tomographic Angiography in Giant-Cell Arteritis. *Stroke* 2018;49:2233-2236
23. Nielsen BD, Hansen IT, Kramer S, et al. Simple dichotomous assessment of cranial artery inflammation by conventional 18F-FDG PET/CT shows high accuracy for the diagnosis of giant cell arteritis: a case-control study. *Eur J Nucl Med Mol Imaging* 2019;46:184-193
24. Koster MJ, Matteson EL, Warrington KJ. Large-vessel giant cell arteritis: diagnosis, monitoring and management. *Rheumatology (Oxford)* 2018;57:ii32-ii42
25. Kermani TA, Diab S, Sreih AG, et al. Arterial lesions in giant cell arteritis: A longitudinal study. *Semin Arthritis Rheum* 2019;48:707-713
26. Espitia O, Blonz G, Urbanski G, et al. Symptomatic aortitis at giant cell arteritis diagnosis: a prognostic factor of aortic event. *Arthritis Res Ther* 2021;23:14
27. Quinn KA, Ahlman MA, Malayeri AA, et al. Comparison of magnetic resonance angiography and (18)F-fluorodeoxyglucose positron emission tomography in large-vessel vasculitis. *Ann Rheum Dis* 2018;77:1165-1171
28. Guggenberger KV, Vogt ML, Song JW, et al. Intraorbital findings in giant cell arteritis on black blood MRI. *European radiology* 2023;33:2529-2535
29. Gospe SM, 3rd, Amrhein TJ, Malinzak MD, et al. Magnetic Resonance Imaging Abnormalities of the Optic Nerve Sheath and Intracranial Internal Carotid Artery in Giant Cell Arteritis. *J Neuroophthalmol* 2021;41:54-59
30. Sommer NN, Treitl KM, Coppenrath E, et al. Three-Dimensional High-Resolution Black-Blood Magnetic Resonance Imaging for Detection of Arteritic Anterior Ischemic Optic Neuropathy in Patients With Giant Cell Arteritis. *Investigative radiology* 2018;53:698-704
31. Mohammed-Brahim N, Clavel G, Charbonneau F, et al. Three Tesla 3D High-Resolution Vessel Wall MRI of the Orbit may Differentiate Arteritic From Nonarteritic Anterior Ischemic Optic Neuropathy. *Investigative radiology* 2019;54:712-718
32. Remond P, Attye A, Lecler A, et al. The Central Bright Spot Sign: A Potential New MR Imaging Sign for the Early Diagnosis of Anterior Ischemic Optic Neuropathy due to Giant Cell Arteritis. *AJNR Am J Neuroradiol* 2017;38:1411-1415

33. Therkildsen P, de Thurah A, Hansen IT, et al. Giant cell arteritis: A nationwide, population-based cohort study on incidence, diagnostic imaging, and glucocorticoid treatment. *Semin Arthritis Rheum* 2021;51:360-366
34. Reichenbach S, Adler S, Bonel H, et al. Magnetic resonance angiography in giant cell arteritis: results of a randomized controlled trial of tocilizumab in giant cell arteritis. *Rheumatology (Oxford)* 2018;57:982-986
35. Seitz L, Christ L, Lotscher F, et al. Quantitative ultrasound to monitor the vascular response to tocilizumab in giant cell arteritis. *Rheumatology (Oxford)* 2021;60:5052-5059
36. Moreel L, Betrains A, Doumen M, et al. Diagnostic yield of combined cranial and large vessel PET/CT, ultrasound and MRI in giant cell arteritis: A systematic review and meta-analysis. *Autoimmun Rev* 2023;22:103355
37. Ing EB, Miller NR, Nguyen A, et al. Neural network and logistic regression diagnostic prediction models for giant cell arteritis: development and validation. *Clin Ophthalmol* 2019;13:421-430
38. Roncato C, Perez L, Brochet-Guegan A, et al. Colour Doppler ultrasound of temporal arteries for the diagnosis of giant cell arteritis: a multicentre deep learning study. *Clin Exp Rheumatol* 2020;38 Suppl 124:120-125

SUPPLEMENTAL FILES



Suppl fig 1: Whole body FDG-PET CT in an 81-year-old with known giant cell arteritis with medium and large vessel (extracranial) involvement. There is increased tracer activity involving the origin of great vessels (B), aortic arch (C), and thoracic aorta (D). Findings are most notable in the bilateral subclavian/axillary arteries (A, black arrows and C, white arrows) (SUV max 2.9). Mildly increased uptake is also noted in bilateral iliac and right femoral arteries (A, arrowheads).



Suppl fig 2: Coronal CTA MIP (A) and VRT (B) images in a patient with LV-GCA show wall thickening and stenosis along the left axillary artery.