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Invited Review

Ultrasound for the Diagnosis of Giant Cell Arteritis

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Abstract

Giant cell arteritis (GCA) is the most frequent large vessel primary vasculitis in the elderly. Correct diagnosis and fast assessment are necessary to prevent complications as well as unnecessary treatments. Giant cell arteritis can present as classical cranial symptoms or as extracranial disease. Although temporal artery biopsy is still the gold standard, ultrasound (US) is gaining ground on evidence with good diagnostic performance as a first approach to support the clinical criteria. The "halo" sign is the most characteristic finding and should be a requisite for reporting an US assessment for GCA with a 43%-77% sensitivity and 89%-100% specificity, when compared to American College of Rheumatology 1990 criteria. Ultrasound is a cost-effective, noninvasive test that offers bed-side results. The need for an experienced sonographer and consensus on the methodology and interpretation of US is fundamental to reduce operator-dependent errors. The diagnostic US algorithm for GCA depends on the clinical scenarios, and in some cases it may be enough to confirm or discard the GCA diagnosis. We review procedure details for cranial and extracranial arteries and technical requirements.

Keywords: Ultrasound, vasculitis, giant cell arteritis, cranial, extracranial, diagnosis

Introduction

Giant cell arteritis (GCA), formerly known as Horton's disease, cranial arteritis, or temporal arteritis, is classified as a large vessel granulomatous vasculitis (LVV). The initial presentation is usually acute, affecting the elderly population.[1] Epidemiological studies show a prevalence in people over 50, with a maximum incidence between 70 and 80. It is more frequent in women than men, with a 3 : 1 proportion. For northern Europe, there is an incidence of 20 for every 100 000 in the risk population.[2] This suggests that gender and genetics are involved in the pathogenesis of GCA.[3]

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Correct diagnosis and quick systemic corticosteroid treatment are necessary to prevent important ischemic complications. American College of Rheumatology (ACR) classification criteria of 1990 (Table 1) require 3 out of 5 criteria,[4] although sensitive, its specificity is given by the histopathologic item. Clinical heterogeneity of GCA can lead to severe complications due to a delay in diagnosis, such as irreversible loss of vision.[5] Classified as a large vessel vasculitis, it can affect vessels of any diameter. Most frequently it affects cranial branches of the external carotid artery, such as temporal and occipital arteries and ophthalmic branch of the internal carotid.[6]

Technical Requirements

The typical signs and symptoms of GCA are associated with cranial arteries' involvement, especially the temporal artery. The most common symptom is temporal headache and mandibular claudication is a very specific sign. Recently, Ponte et al[12] published the frequency of presentation of symptoms in GCA (Table 2). Scalp sensitivity and visual disturbances are frequent, and temporal arteries (TA) may be thickened and pulse decreased or absent. Elevation of inflammatory parameters such as eritrosedimentation rate and C-reactive protein is common. Cranial involvement related to GCA is serious and can lead to ischemic events, including blindness due to ischemic optic neuropathy and strokes. These events can occur in the initial phase of the disease, and for this reason, GCA must be considered a medical emergency.[1] Giant cell arteritis can also present systemic symptoms such as fever, weight loss, and polymyalgia rheumatica (PMR) is frequently associated.[7]

Different phenotypes of the disease have been described, besides the classical cranial GCA, extracranial GCA with involvement of large vessels, mainly the thoracic aorta, with or without PMR is described in up to 30%-50% of patients.[8,9] In the case of extracranial GCA, patients can be asymptomatic or have claudication in the extremities, a vascular murmur, and decreased or absent pulses.[10] Potential and late complications include valvular disease and aortic aneurysms and/or dissections.[11,12]

Table 1. American College of Rheumatology 1990 Criteria for the Classification of Giant Cell Arteritis*

1. Age at disease
2. New headache
3. Temporal artery abnormality such as tenderness to palpation or decreased pulsation
4. Erythrocyte sedimentation rate >50 mm/h
5. Abnormal artery biopsy showing vasculitis with mononuclear cell or granulomatous inflammation, usually with giant cell infiltrates

*For purposes of classification, a patient shall be said to have giant cell arteritis if at least 3 of these 5 criteria are present. The presence of any 3 or more criteria yields a sensitivity of 93.5% and a specificity of 91.2%[4]

Although temporal artery biopsy (TAB) is still the "gold standard," over the last 25 years, the field of imaging and GCA has had a rapid expansion and has brought about controversy over when, which, and whom. Recently, EULAR has developed evidence-based recommendations where an early imaging technique is proposed as a first approach to support the clinical criteria. It aims to shorten time for diagnosis, reduce complications, and avoid unnecessary treatment.[13] While some questions still remain, some general considerations are clear (LoE 1 LoA 90%), temporal +/− axillar artery ultrasound (US) is recommended as the first imaging modality in cranial GCA, assuming high expertise, appropriate equipment, and prompt availability.[13]

Evidence on Ultrasound for Giant Cell Arteritis Diagnosis

Several meta-analyses and systemic literature reviews have assessed the performance of US abnormalities to diagnose GCA. When compared to ACR criteria, US halo sign has a sensitivity of 43%-77% and specificity of 89%-100%.[14-18] We have also learned that the test's sensitivity and specificity can be increased if the halo sign is present at more than one artery, up to 100% if bilateral.[19,20] The compression sign, later detailed, has shown good diagnostic performance, not only because of its simplicity and high interobserver agreement (0.92), but because it is comparable to the halo sign in daily practice with up to 100% positive predictive value for GCA diagnosis.[21,22]

The TABUL (Temporal Artery Biopsy and Ultrasound in diagnosis of Giant Cell Arteritis) study[23] is a prospective, multicenter study of new cases, comparing US and TAB to clinical diagnosis. The objective was to evaluate the diagnostic performance of US as an alternative to biopsy for GCA diagnosis and to compare its cost-effectiveness. The sensitivity and specificity of US when compared with reference diagnosis was 54% and 81%, respectively, meanwhile for TAB was 39% vs. 100%. A combination strategy using both modalities, with TAB in those patients with negative US, increased the sensitivity to 65%. Cost-effectiveness leaned toward US.

We must keep in mind that an operator-dependent error may limit its facticity. A learning curve of 300 explorations is recommended to rely on the results[12,24] so it is important to remember that false–positive or false–negative tests may appear (Table 3). Atherosclerosis can mimic the halo sign,[25] and other diseases can present a halo sign due to inflammatory infiltrates, which lead to wall thickening and translate to a hypoechoic halo around the arterial lumen.[24] The halo sign has been found in 4.6% of tests as a false–positive for GCA in other diseases such as systemic vasculitis (anti-neutrophilic cytoplasmic antibody-associated vasculitis, polyarteritis nodosa), infectious secondary vasculitis, or vasculitis like (osteomyelitis of the skull base, neurosyphilis), hematological conditions (non-Hodgkin lymphoma and multiple myeloma).

We must remember that the performance of TA +/− axillary arteries (AA) US depends on the clinical situation where it is applied and will show the highest rentability in those who present with concordant clinical data. Nevertheless, it seems that the more branches are affected, the higher the diagnostic yield of the test.[15,19,23,24]

Temporal Artery Biopsy Versus Ultrasound for Giant Cell Arteritis

An open debate still exists on whether US is better than TAB, and to date, no consensus exists as different factors fall into place. Assessing the diagnostic performance of TAB has some limitations in GCA since different histological patterns of inflammatory infiltrates on TAB have been found.[26] The heterogeneity of the disease, the skip lesions, its distinct phenotypes, the influence of previous treatments, and sampling, all influence this controversial issue. Rinagel et al,[23] in a recent meta-analysis, reported a sensitivity of 68% and a specificity of 81% of the hypoechoic halo compared to positive TAB. The TABUL study carried out an interrater analysis for biopsy results with variability among pathologists (0.62 correlation coefficient, CI, 0.49-0.76). The TABUL study found a sensitivity of 39% with a 100% specificity. Other options with higher sensitivity could improve patient care in GCA.[12]

Table 2. Frequency in Presentation of Symptoms in Giant Cell Arteritis

Clinical Feature	Frequency (%)
Elevate erythrocyte sedimentation rate and/or elevated C-reactive protein	90-95
Headache	70-90
Polymyalgia rheumatica	40-60
Constitutional symptoms (low-grade fever, fatigue, or weight loss)	30-60
Abnormal temporal artery on physical examination (tenderness or absent or diminished pulses)	30-60
Jaw claudication	40-50
Scalp tenderness	33-50
Visual disturbances (transient or permanent)	20-50
Cerebrovascular accident (transient ischemic attack or stroke)	3-7

*Adapted from C Ponte (2020).[7]

Table 3. Challenges for the Halo Sign

Problem	Solution	Examples
False positive (specificity)	Look for more vascular territories (quantity of halos) Look for high cutoff points (quality of halos)	Atherosclerosis Other vasculitis Infection Malignancy
False negative (sensibility)	Learning curve	Operator dependent Incorrect probe Technique pitfalls

The Sooner the Better

Diagnostic delay is still a challenge in GCA. Recent publications confirm that the prevalence of visual loss still represents 25% of patients.[27] Rheumatologist-led fast track diagnosis and management pathway for patients with possible GCA has proven to reduce sight loss and morbidity.[28-30] It must coordinate a "fast" assessment, which includes clinical, laboratory, and TA and axillary artery US assessment. Support from an ophthalmologist and surgeon, as well as communication with the general practitioner, emergency room colleagues, and from other specialties, is also needed.

Ultrasound should be performed as early as possible, best within 1 week of starting glucocorticoid therapy because it could reduce the sensitivity of imaging. Treatment should never be delayed while awaiting the test.[13]

Pros and Cons of Temporal Artery Ultrasound

The performance of a diagnostic test depends on its sensitivity and specificity and on the clinical situation where it is applied. For GCA, this pretest probability score is not yet available and must be determined case by case based on clinical probability.[13] Ultrasound should be the first imaging test in patients with suspected GCA presenting with predominant cranial symptoms (Table 4).

For patients with LVV-GCA, the aorta cannot be correctly assessed with US, and the imaging test to be performed should be selected based on availability and expertise. Still, a rapid scan of axillary and supraaortic vessels can be done. The EULAR task force accorded that if GCA cannot be confirmed or excluded based on clinical, laboratory, and imaging results, TAB and/or additional imaging is required.[13]

Proposed Algorithm

The diagnostic algorithm for GCA depends on the clinical presentation, the available imaging methods, and the local expertise.[31] A positive US and a high clinical suspicion are probably enough to make the diagnosis as well as a negative US with a low probability for the GCA diagnosis. The question remains for those in between, with inconclusive US or unspecific symptoms. In this respect, alongside and in the same direction, algorithms for GCA diagnosis have been published[12,32,33] where complementary tests are needed (Figure 1).

Future Milestones on Ultrasound for Giant Cell Arteritis

A publication is expected soon for an update on diagnostic and classification criteria for vasculitis, based on a large multinational, observational study DCVAS (Diagnostic and Classification Criteria in Vasculitis Study). So far, Ponte et al[34] have made a draft classification criteria for LVV (GCA and Takayasu), which includes different weighted criteria for clinical features, acute phase reactants, TAB, and specific imaging findings (including the halo US sign) which report good sensitivity and specificity in the validation cohorts, 89% and 91%, respectively.

Recently, van der Geest et al[35] have proposed a US halo score for GCA assessment to quantify the extent of vascular inflammation that may help to discriminate between patients with high (>30%) or low (<5%) risk for ocular ischemia. The grading for this score comes from measurements of halo thickness in different segments at TA and AA.

As we wait for validation on these novel proposals, the US is paving perspective and seems to be enough to confirm or discard GCA in certain clinical scenarios.

As we have commented previously, various studies demonstrate the high sensitivity and specificity of US in the diagnosis and management of large vessel vasculitis. They all have 2 common features: 1) the importance of an experienced sonographer and 2) the need for consensus on the methodology and interpretation of the US technique. Shown below, we proceed to detail technical descriptions for US diagnosis in GCA.

Vascular Ultrasound Imaging of Temporal and Extracranial Large Arteries: Technique Skills

As discussed in previous sections, US in the TA is particularly valuable for the diagnosis of GCA.

Table 4. Pros and Cons of temporal artery ultrasound (TAUS)

Pro	Cons
Noninvasive, nonradiating, fast, bedside results	Operator dependent
Cost-effective	Learning curve
Good diagnostic performance (sensitivity and specificity)	No imaging of thoracic aorta

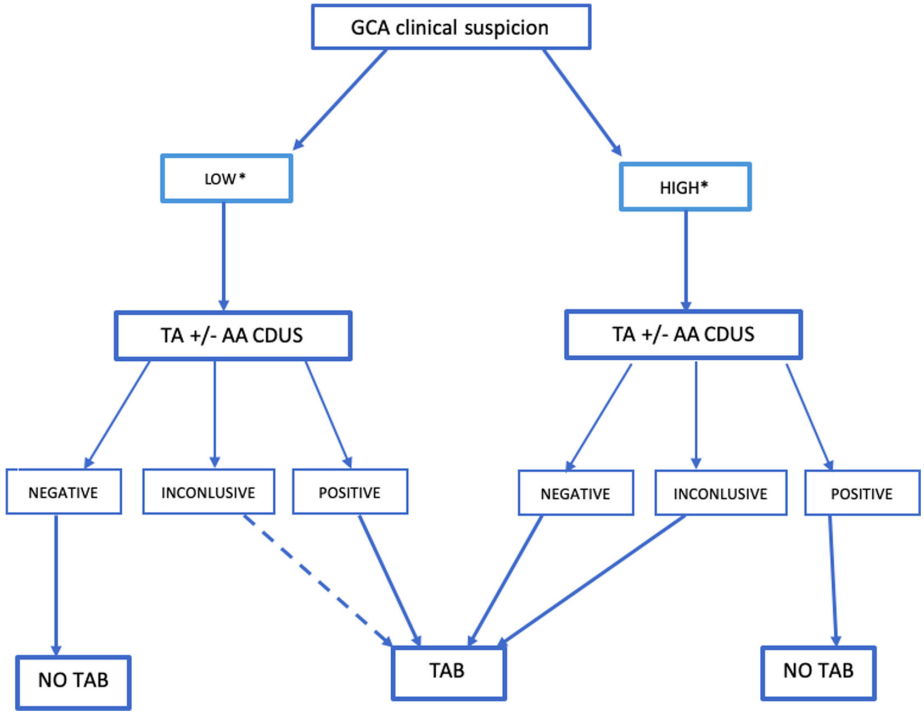

Figure 1. Proposed algorithm for the diagnosis of GCA. *High clinical suspicion: age >50 years and 1) cranial symptoms (new onset headache, jaw claudication, or visual symptoms); 2) polymyalgia rheumatica according to European Alliance of Associations for Rheumatology (EULAR)/polymyalgia rheumatica 2017 criteria; 3) toxic syndrome or fever from unknown origin, once infectious and neoplastic causes are ruled out; 4) vertebrobasilar stroke, without cardiovascular risk factor history for atherogenic or embolic etiology. AA, axillary artery; GCA, giant cell arteritis; TA, temporal artery; TAB, temporal artery biopsy. Adapted from Corominaset al (2019).[32]

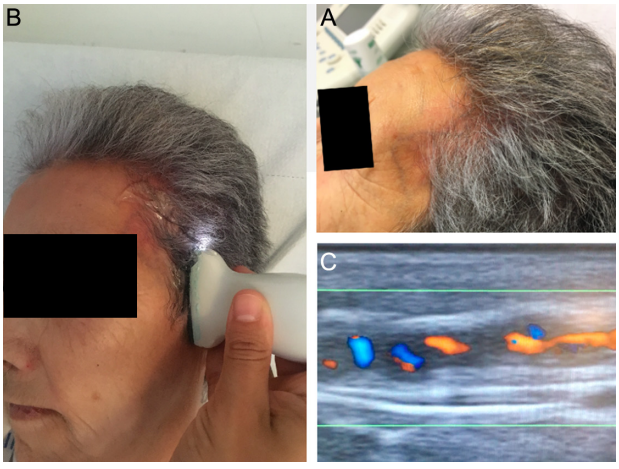

Figure 2. Temporal artery ultrasound exploration. A: The left temporal artery looks thickened and erythematous. B: Probe on longitudinal scan of the parietal branch of the temporal artery. C: Halo sign and stenosis on longitudinal scan.

To obtain the most reliable examination, common superficial TA with their frontal and parietal branches should be examined (Figure 2).

It has also been demonstrated that examination of the AA increases the sensitivity of US in the diagnosis of GCA. It has been reported that 50% of GCA patients could have axillary artery involvement.[7,8] In 2008, Schmidt et al[7] published a study whose aim was to describe characteristic US findings of patients with newly diagnosed GCA. They concluded that performing axillary artery US in all patients with suspected temporal arteritis increases the diagnostic yield for LVV-GCA. In fact, in 2014, Schmidt et al[7] described that an experienced sonographer examining both the temporal and AA could find more GCA patients than a biopsy with histology of the common superficial TA.[36] Therefore, EULAR 2018 recommendations for the use of imaging in LVV in clinical practice recommend US of both the TA and AA as the first imaging modality examination to be performed in all patients with suspected GCA and cranial manifestations. Furthermore, the AA can be easily assessed as long as the patient has no severe limitation in shoulder mobility.

In recent years, thanks to new diagnostic tools, a high frequency of extracranial disease involvement in GCA patients has been described, motivating different research teams to study US findings in extracranial territories apart from AA, as occipital, facial, carotid, and vertebral arteries in patients with GCA.[7,37]

Once again, occipital arteries are assessed by US because they are located superficially posterior to the ear. These arteries are affected less frequently than TAor AA, but they are specifically affected in some patients, particularly if they present with retroauricular pain.[38] Facial arteries are also are assessed by US, as they wind around the body of the mandible. Ultrasound detects facial artery involvement in 31% of GCA patients, but this percentage can increase in patients with visual impairment or jaw claudication.[36,39]

Diamantopoulos et al[30] in 2014 described that adding the exploration of the common carotid artery had an excellent sensitivity and high specificity to diagnose GCA.[30] When concerned about carotids and GCA, 2 aspects should be taken into account: 1) the common carotid arteries are more often involved than the proximal internal and external carotid arteries in GCA, and 2) arteriosclerosis of carotid arteries is common among the age group of patients with suspected GCA.

Another of the territories that has been proposed to study by the US in patients with GCA are the vertebral arteries. The vertebral arteries can also be easily evaluated by US. The probe allows us to objectify segments of the arteries between the vertebral transverse processes. Vasculitis involvement of the cerebral arteries should be suspected in patients with cerebral infarcts and elevated acute phase reactants because the involvement of the vertebral arteries usually occurs with infarcts in the affected territory.[40] Ultrasound allows us to evaluate the subclavian arteries at the mid and distal level. The proximal part of the subclavian artery and the left common carotid artery can only be seen at a lower resolution because they are located deeper. Therefore, routinely examining these arteries may not increase the sensitivity of US greatly, as they are less commonly involved than axillary and/or TA.[7,8,36]

If the US examination of the temporal and AA is inconclusive and the clinical history and complementary examinations are suggestive of GCA, an examination of other vessels such as the aorta can be performed, but with less sensitivity than those previously described.[37] Examination of the thoracic aorta by US is not of value due to overlapping of the lungs. Only the exploration of its first 4 cm and with low-frequency probes would be valuable in the case of severe pathology. Visibility of the abdominal aorta is generally better with US, although the resolution is low in obese patients. It should be noted that meteorism can further decrease image quality.

It is known that the involvement of the superficial femoral arteries and popliteal arteries is commonly seen in GCA.[8] Ultrasonography of the lower extremities allows us to study arterial occlusive disease, whether it is atherosclerotic (the most frequent) or of vasculitic etiology. In patients with suspected GCA, the pulse of the pedal arteries should be taken. If absent, the US can help us differentiate if the occlusions are due to atherosclerosis or vasculitis.

Sonographic and Clinical Pattern of Extracranial and Cranial Giant Cell Arteritis

An Outcome Measures in Rheumatology (OMERACT) Large Vessel Vasculitis Ultrasound Working Group was constituted to standardize definitions of the elementary US lesions suggestive of GCA.[25] They intend to reach consensus for definitions of normal and pathological GCA vascular characteristics that appear on temporal and extracranial large arteries as detected by US.

Before focusing on the vasculitis US images that can be seen in patients with GCA and LVV, the sonographer should feel comfortable with the exploration of normal arteries. Nonpathological TA are characterized by pulsating imaging, a compressible artery with an anechoic lumen surrounded by mid-echoic to hyperechoic tissue. The intima–media complex is a homogeneous, hypoechoic, or anechoic echostructure delimited by 2 parallel hyperechoic margins (described as "double line pattern") (Figure 3).

Besides, extracranial large arteries in nonpathological conditions are also characterized by pulsating images, a hardly compressible artery with an anechoic lumen. The intima–media complex is presented in the same way as in the TA, with a double-line pattern. All examinations should be performed in longitudinal and cross sections.

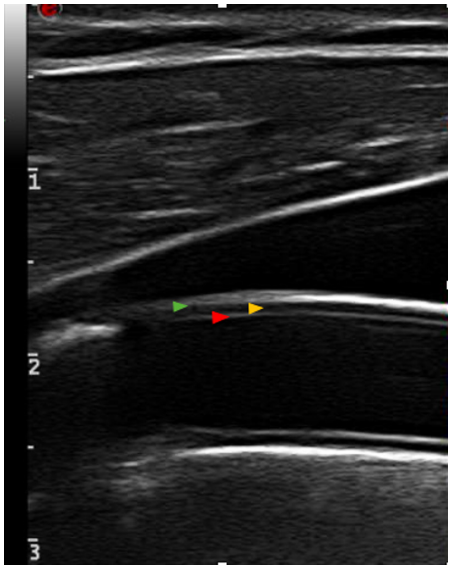

Figure 3. Nonpathologic axillary artery ultrasound. Longitudinal normal section. Pulse repetition frequency at 2.5. The intima (red arrowhead), media (yellow arrowhead), and adventitia (green arrowhead) can be visualized.

The US findings described in patients diagnosed with GCA are as follows:

- Halo sign: The halo sign is the most characteristic US finding of the GCA (giant cell arteritis). It was first described in 1995. Inflammation of the vascular wall translates sonographically into homogeneous hypoechoic thickening of the wall. It is a well-delineated image toward the luminal side and should be visible in both longitudinal and transverse planes, although it is more commonly observed in transverse concentric planes[7] (Figure 4, 5).

The EULAR working group considered the halo sign to be the most important US sign for cranial and extracranial GCA with 100% and 83.3% agreement, respectively. In fact, many publications suggest that the halo sign should be a requisite for reporting a US assessment as a vasculitis diagnostic.

The role of intima–media thickness (IMT) measurements for diagnosis and monitoring of ACG is still uncertain. Several studies have proposed different cutoff values, but none of them have been validated. Therefore, the definition of the halo sign proposed by the OMERACT group does not include measurement of IMT.

The range of cutoff values for the halo sign, proposed for TA is 0.3-1 mm and for large extracranial arteries is 1.3-2 mm.[7,41-47]

- Stenosis: According to the OMERACT definition,[25] stenosis in the TA is characterized by aliasing and persistent diastolic flow by color Doppler US (Figure 6). The maximum systolic flow velocity determined within the stenosis by pulsed wave-Doppler US is ≥2 times higher than the flow velocity proximal or distal to the stenosis. In extracranial large arteries, US images show a typical vasculitic vessel wall thickening with characteristic Doppler curves showing turbulence and increased systolic and diastolic blood flow velocities.
- Occlusion: Occlusion is defined as the absence of color Doppler signals in a visible artery filled with hypoechoic material, even with low pulse repetition frequency and high color gain.
- Compression sign: The compression sign should be assessed by applying pressure via the transducer until the lumen of the temporal artery occludes and no arterial pulsation remains visible. In the healthy artery, the lumen disappears upon compression; for the pathological vasculitic artery, this sign makes reference to the persistence of the hypoechogenic image of vessel wall thickening.[21]

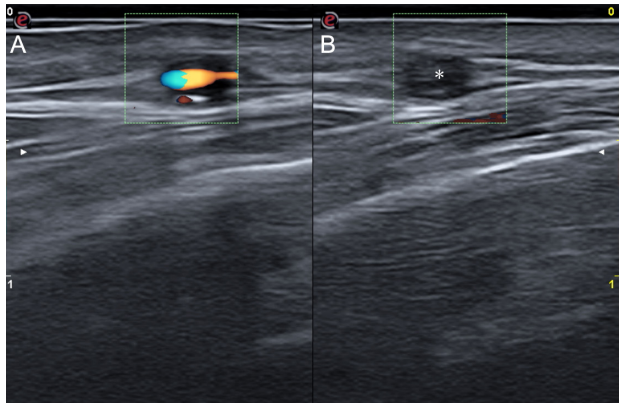

Figure 4. Halo and compression signs. A: The halo sign for the temporal artery on a short scan. B: Positive compression sign, where the hypoechoic area persists (asterisk) during the compression maneuver of the vessel lumen with the probe.

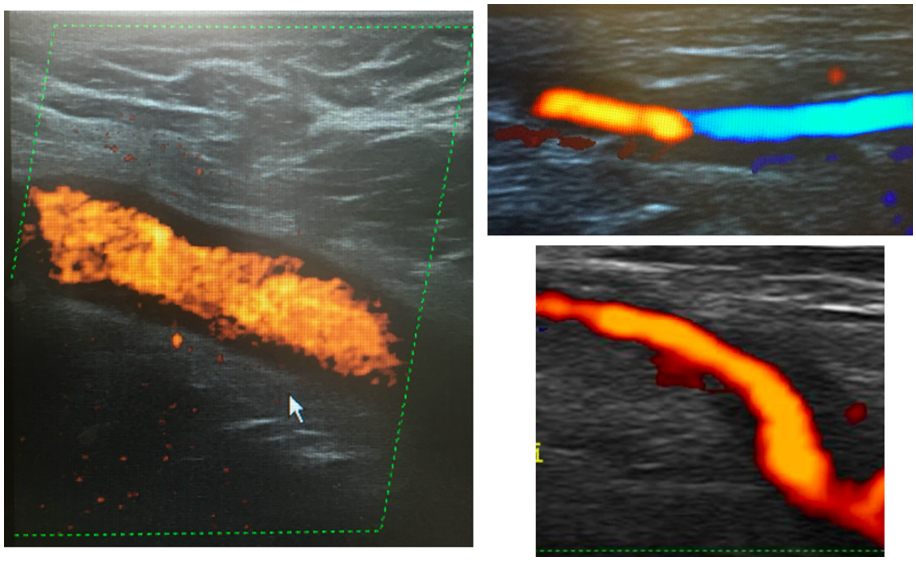

Figure 5. Halo sign. The halo sign (arrow) is seen as homogeneous hypoechoic wall thickening toward the luminous side. A: Longitudinal scan of the axillary artery. B: The frontal branch of temporal artery. C: The parietal branch of temporal artery.

Technical Requirements

Depending on the vascular territory to be explored, US examination should be performed by a trained specialist using the appropriate equipment, established operating procedures, and appropriate settings. The US equipment that we use for musculoskeletal system explorations in our routine practice is usually apt for vascular exploration of both the temporal and extracranial arteries. For supra-aortic arteries, linear probes are recommended; for the ascending aorta, aortic arch, and abdominal aorta convex probes.

As in all US examinations, depending on the depth of the vascular territory to be explored,

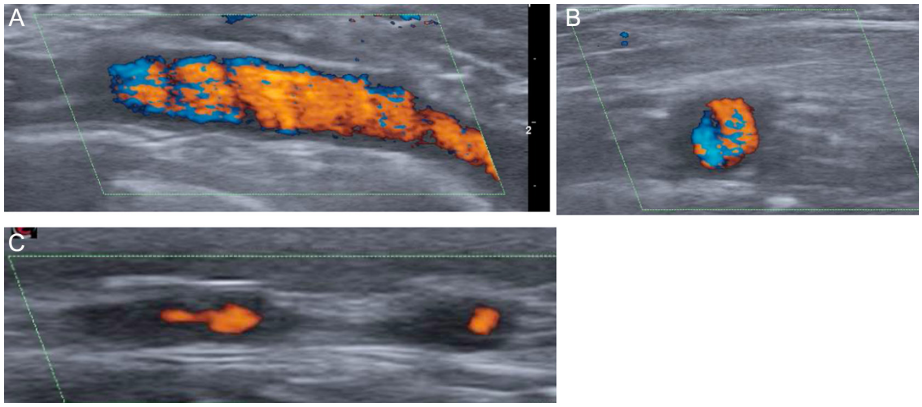

Figure 6. Ultrasound of the temporal artery of a patient with active ACG. Halo sign (arrowhead) on longitudinal (A) and trans(B) scan. Stenosis (line) of the temporal artery (C).

different B-mode frequencies will be applied. For superficial arteries, high frequencies as ≥15 MHz are required in order to detect minor wall thickening and provide higher resolution. Image depth should be 10-20 mm for TA. To explore deeper arteries such as extracranial supra-aortic arteries, low frequencies are required as 7-15 MHz in order to gain more penetration. Image depth should be 30-40 mm for extracranial supra-aortic arteries.

The focus should be at the level of the artery we are exploring. To avoid the anechoic appearance of the artery wall and the underfilling or overfilling of the vessel lumen, the B-mode gain and the color Doppler gain should be adjusted.

EULAR 2018 recommendations of the images in LVV[13] recommended color Doppler mode over power Doppler mode. The application of Doppler in LVV is very different from the one for arthritic conditions. For the TA, Doppler frequencies should be of 7-12 MHz and for the extracranial supra-aortic arteries of 4-8 MHz. Pulse repetition frequency should be 2-3.5 kHz and 3-4 kHz, respectively. The angle between sound waves and the artery should be ≤60°. Pulse repetition frequency correct adjustment is crucial in order to avoid aliasing and motion artefacts.[48] There are Doppler artifacts that we have to know in order to recognize them, like mirror and reverberation artifacts.

Conclusion

Giant cell arteritis requires a correct diagnosis and quick systemic corticosteroid treatment to prevent important ischemic complications and avoid unnecessary treatments. Although TAB remains the "gold standard" for diagnosing GCA, imaging techniques have gained relevance in the diagnosis and management of GCA. High sensitivity and specificity of US of the TA and AA have been described in the diagnosis of GCA. In this scenario, EULAR has developed evidence-based recommendations proposing the use of US as the first imaging modality in cranial GCA. The main US findings observed in ACG is the halo sign and compression sign. The TA and AA have shown the most profitability in the diagnosis, but patients with extracranial symptoms may benefit from US examination of other vascular territories. Therefore, vascular US helps us in the diagnosis of GCA. Due to its easy accessibility and its high sensitivity and specificity, it could be considered the main test for the diagnosis of all patients with GCA and cranial manifestations and some patients with GCA and extracranial manifestations.

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