

Guidance for Giant Cell Arteritis Ultrasound and Service Provision

BMUS 

RECOMMENDED PRACTICE GUIDELINES

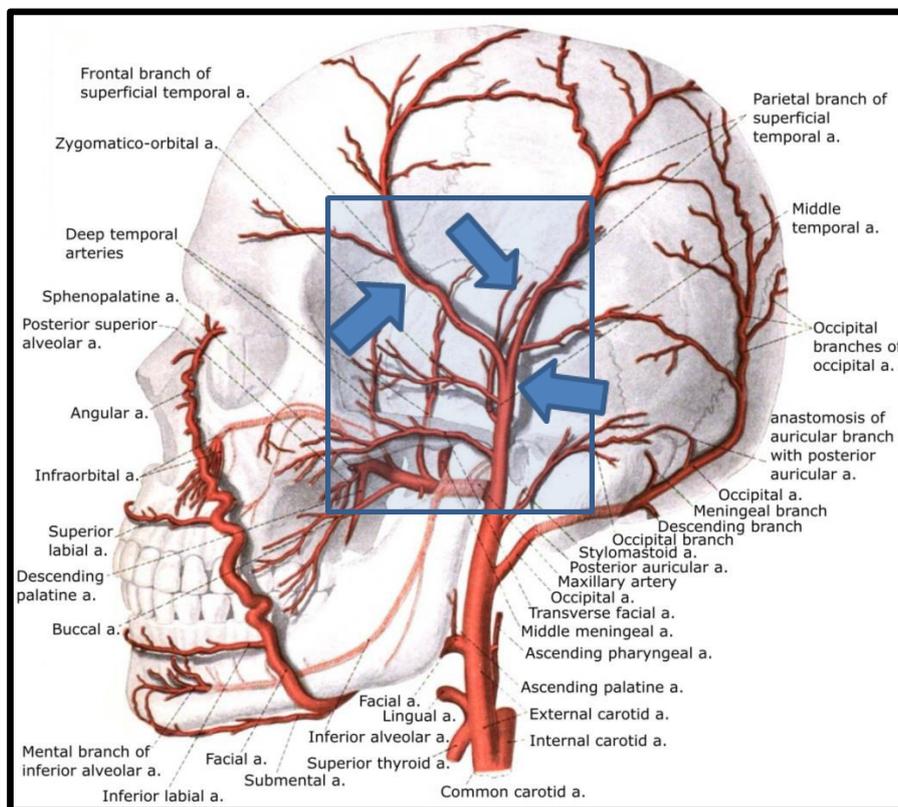
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1. What is GCA?

Giant cell (or temporal) arteritis (GCA) is the most common forms of systemic vasculitis in adults over the age of 50 years. It primarily affects medium and large vessels and is most evident in the temporal and axillary arteries of adults. Its clinical presentation includes headache often with severe pain over the temples and scalp, jaw pain when chewing, or polymyalgia rheumatica-like symptoms; flu like symptoms such as fever and malaise. In its most severe form, it can cause uni or bilateral vision loss secondary to arteritic ischemic optic neuropathy. GCA is considered a medical emergency and its prompt diagnosis and treatment are essential to avoid blindness.

In most hospital Trusts, presenting patients are referred to the rheumatology department for the diagnosis and treatment of GCA and there is increasing demand for rapid access to diagnostic pathways that include ultrasound.



By DR. Johannes Sobotta. Updated nomenclature by Mikael Häggström - Original publication: Atlas and Text-book of Human Anatomy Volume III Vascular System, Lymphatic system, Nervous system and Sense Organs. 1909., Public Domain,
<https://commons.wikimedia.org/w/index.php?curid=57573335>

2. Diagnostic Imaging for GCA

Temporal artery biopsy is deemed the gold standard for diagnosis, however 15% or more patients with GCA are reported to have a negative biopsy result despite adequate tissue sample (1).

The British Society of Rheumatology (BSR) and the European League against Rheumatism (EULAR) both now recommend Ultrasound as a first line investigation for GCA.

Imaging should be performed by a trained specialist using appropriate equipment, operational procedures and settings (2).

Imaging should be performed before or as early as possible after initiation of therapy, best within 1 week, as treatment with glucocorticoids rapidly reduces the sensitivity of imaging.

- In patients with suspected GCA an early imaging test is recommended to complement clinical criteria. It is necessary to assess for the presence of active inflammation in the temporal and axillary arteries. The presence of a 'halo' sign is highly diagnostic of GCA (describe later in the text)
- In patients with a high clinical suspicion, if the imaging is positive a biopsy is not required
- In patients with a low clinical suspicion, with negative imaging, GCA can be considered unlikely
- Ultrasound, PET, MRI and /or CT may be used to detect mural inflammation in extra-cranial arteries to support diagnosis of LV-GCA.

In writing this guidance, a presumption is made that all examinations will be carried out by trained practitioner with competencies signed off by a qualified supervising practitioner and acts according to BMUS Guidelines for Professional Practice (3).

3. Referral System and Diagnostic Pathway

Ultrasound referrals for patients with suspected GCA are urgent and should only be made by referring Rheumatology specialists following specialist review in a rapid access clinic.

Referrals from Primary Care and in most cases other specialities are not appropriate and a robust referral system is advised.

Any service provided would otherwise risk being overwhelmed by low-risk referrals for headache. Also, a negative scan does not always mean that the patient does not have GCA, so appropriate management on clinical grounds is vital.

Initial treatment will include high dose oral steroids which, will reduce or even resolve the ultrasound appearances of GCA. It is therefore recommended that Imaging should be performed before or as early as possible after initiation of therapy, best within 1 week (2)

Prior to starting a GCA service, it is important to meet and agree diagnostic pathways and referral criteria with your Rheumatology Team (5). Due to the urgent nature of these referrals patients should be seen by the referring specialist following the scan. Arrangement should ideally be made with the Rheumatology team for same day clinical review with scan findings.

Common Indications for Referral Include:

- Acute Onset (symptoms <6weeks)
- Abnormal CRP and ESR
- Age >50
- Ocular / Ophthalmic Symptoms - transient visual loss, diplopia, visual fatigue or visual field loss

i.e. indicating CRAO (central retinal artery occlusion) or AION (acute ischaemic optic neuropathy)

- Tender, thickened, palpable or pulseless temporal artery (TA)
- Scalp Tenderness or Scalp Necrosis
- Jaw or Tongue Claudication (including difficulty opening mouth)
- Constitutional - weight loss, loss of appetite, sweats, fatigue, malaise
- Sudden onset arm claudication

Referrals to Decline / Exclude

(unless specifically requested after Consultant assessment from your Rheumatology Team)

- Normal bloods
- Vague headaches
- Age <50
- Length of symptoms >6weeks

Essential Information Required

A report proforma may be used (see sample provided) making sure any electronic request system mandates the collection of the following information as this is crucial to interpreting the scan results

- Date of Onset of Symptoms
- Blood results
- Date of Referral
- Whether On Steroids - Yes / No
- Date of Initiation of Steroids
- Whether Previous Temporal Artery Biopsy (TAB) - Yes / No

Appointment Times for GCA Ultrasound Examination

30min is advised - longer may be necessary initially until a level of experience is achieved across all operators.

There should be adequate numbers of trained staff to ensure robust service provision.

4. Technique

Patient Positioning and Preparation

It is usual practice for the patient to be supine or semi-recumbent, so that the practitioner is not overreaching to assess the frontal branches. The patient's head may be supported by a pillow to ensure neck support. Resting a cushion on the patient's chest can help reduce pressure on the patient and aid as an arm rest to steady the practitioner's hand.



Loose clothing or a gown to ensure ease of access to the chest/axillary area is recommended. No specific preparation is required for the examination, however, if the patient has been commenced on steroids, length of time in days must be noted in the report.

Equipment and Settings

A linear high frequency transducer is ideal to assess the superficial temporal arteries (STA) due to the small size of the vessels, for example a 15-18MHz Hockey stick transducer.

Assessment of the axillary arteries is better suited to a medium/high frequency transducer.

A vascular preset, optimised for GCA scanning (small vessels with low flow) by your applications specialist according to BSR and EULAR guidance, is advised. See further references for guidance on equipment settings (6,7)

British Society of Rheumatology – B Mode Settings

SETTING	TEMPORAL	AXILLARY
Frequency	Highest possible >15MHz	10-15MHz
Depth	1-2cm, then adjust to patient habitus	3-4cm, then adjust to patient habitus
Focus	0.5cm, then adjust to the level of the artery	2cm, then adjust to the level of the artery
Gain	35-45dB, adjust so halo is hypoechoic, not anechoic	35-45dB, adjust so the halo is hypoechoic, not anechoic
Make sure the halo is visible in two planes and the compression test is positive.		

British Society of Rheumatology – Doppler Settings

SETTING	TEMPORAL	AXILLARY
Mode	Colour (consider Power Doppler for very slow flow)	Colour
Steer of Box	<60degrees towards flow	<60degree towards flow
Size of Box	Not too tall or wide, enough to cover the lumen	Not too tall or wide, enough to cover the lumen
PRF	2-3.5 kHz	3-4 kHz
Frequency	7-12.5 MHz	5-8 MHz
Gain	Enough to fill the lumen and avoid blooming	Enough to fill the lumen and avoid blooming

These tables are a useful reference point but not be absolute.

5. Scan Protocol

During the examination the ultrasound practitioner should examine the following:

The common, parietal and frontal branches of the temporal arteries in both B-mode and colour/power Doppler in longitudinal and transverse.

- Turn the patients head away or toward you to access the temporal region.
- Locate the common branch of the STA in transverse by the ear/parotid, where the vessel is at its largest.
- Once you have located the STA in transverse, slowly rotate the probe into longitudinal orientation to assess the rest of the vessel in B mode and colour.
- Follow the STA caudally in transverse until you reach the bifurcation.
- Follow the frontal and parietal branches as far as possible, using colour/power Doppler and compression to assess the vessel.
- Patients will often demonstrate a region of focal tenderness, which is always worth rescanning at the end of the exam.
- Where there are signs of wall thickening (reduced luminal size with circumferentially thickened walls) confirm this by compressing the vessel with the probe; a non-diseased vessel will fully compress leaving no residual vessel behind, diseased vessels with positive halo will have circumferentially thickened walls that do not compress (Halo sign).

The halo should be present in two planes and be circumferential¹⁰. Inflammation can cause occlusion, stenosis or may not be flow limiting⁹. The Halo thickness should be >0.3mm⁹, moreover, a halo thickness of 0.7mm or greater can predict a positive biopsy result¹¹.

Examination of the axillary arteries should also be performed.

- It is recommended to start with anterior view (with arm resting by the patient's side with some bend in elbow) and then if necessary have patient raise arm to rest behind them on pillow allowing access to the Axilla. Locate the axillary artery in transverse in the axilla and examine using colour/power Doppler.
- Turn the transducer through 90 degrees and locate the head of humerus. Elevate the probe angle towards the ventral aspect of the patient anteriorly and heel toe the probe to bring the vessel parallel to the probe to obtain good specular reflection from the vessel wall. Again using colour/power Doppler. Compression will not be possible with such a large artery.

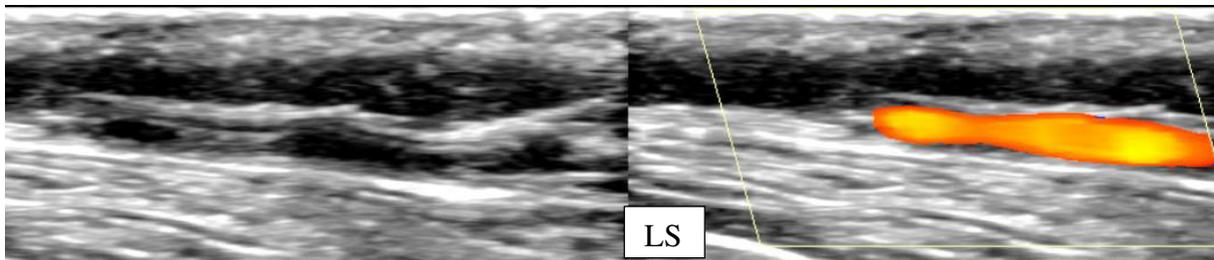
In both anatomical areas:

- Adequate machine setting manipulation must be employed to avoid / prevent over or under filling of the vessel on colour/power Doppler.
- Any intimal thickening should be identified and measured and its approximate location/extent recorded in the report (see below).

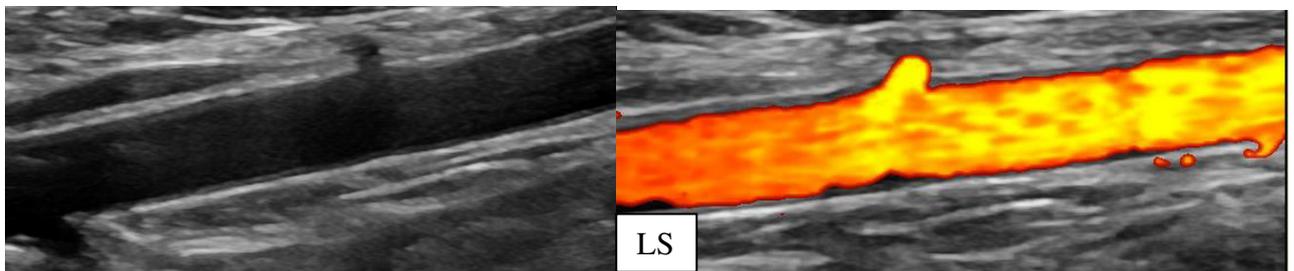
6. Ultrasound appearances of normal vessels and in GCA

On ultrasound, a normal artery appears as:

- Pulsating and compressible with anechoic lumen surrounded by mid-echoic to hyperechoic tissue
- With high resolution equipment, the intima-media complex is seen as homogeneous, anechoic echo structure delineated by two parallel hyperechoic margins “double line pattern” may be visible – see below (8).



Grey scale and Doppler images of a normal temporal artery

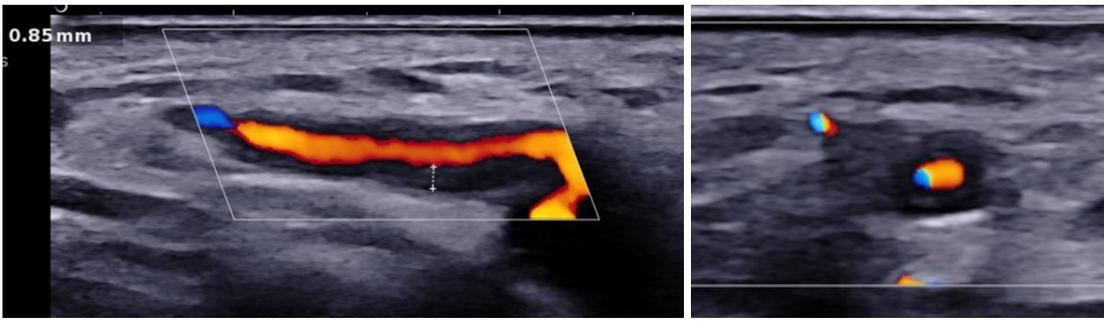


Grey scale and Doppler images of a normal axillary artery

7. Features of Giant Cell Arteritis

‘Halo’ Sign

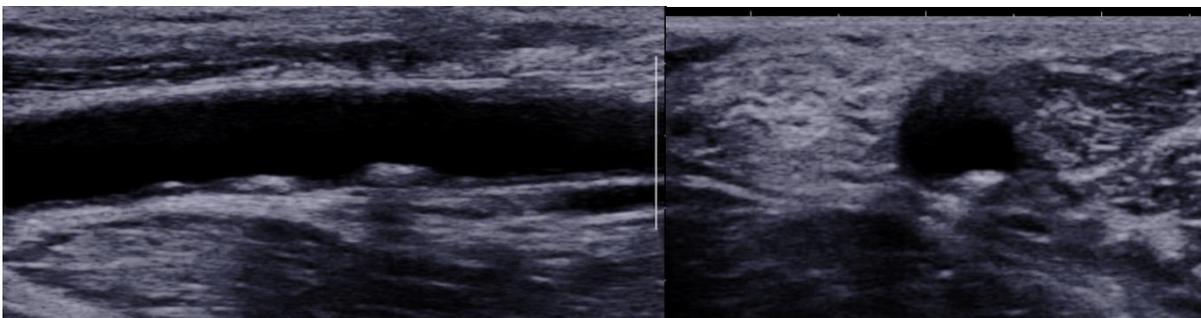
- Homogeneous and hypoechoic vessel wall thickening – commonly called the ‘halo’ sign
- Well delineated - can be seen on B-mode, colour or power Doppler or with B-flow imaging (below)
- Usually symmetrical - concentric vessel wall alteration/stenosis that can result in occlusion of the artery.
- The narrowed lumen can be imaged with colour/power Doppler modes - spectral Doppler is rarely necessary as this only describes what the eye can see and there are no treatment criteria based on measuring the degree of stenosis



Doppler ultrasound image of a temporal artery showing hypoechoic, concentric wall thickening in a patient with GCA.

Atherosclerosis

An alternative pathology causing arterial wall thickening is atherosclerosis. Unlike GCA, these deposits are heterogeneous and in part hyperechoic, irregular delineated and eccentric vessel wall alteration.



Ultrasound image of an artery in a patient with atherosclerosis

Halo Sign - Single Wall IMT Measurement

Identification of any intimal thickening should be carefully measured and the maximum thickness recorded in the report if it exceeds the normal range of cut-off values, stating whether this is related to GCA/large vessel vasculitis (LVV) or atherosclerosis.



Ultrasound images with measurements of abnormal wall thickening in a patient with GCA.

The image may need to be magnified before measuring the wall thickness or IMT to avoid measurement errors. The posterior wall of the axillary artery is the normal site for measurement of the IMT to avoid anterior wall reverberation artefact.

These are the suggested cut off values for normal, versus vasculitis in patients with GCA (9).

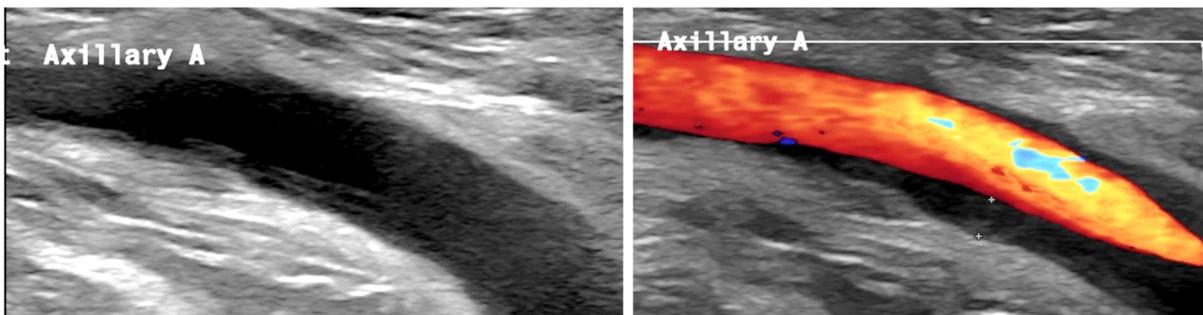
Halo Sign – Cut Off Values

Common Temporal Artery	0.42
Frontal Artery	0.34
Parietal Branch	0.29
Axillary Artery	1.0

These measurements are highly dependent on excellent technique, operator presets and accurate equipment manipulation and therefore should be used with a degree of care and caution.

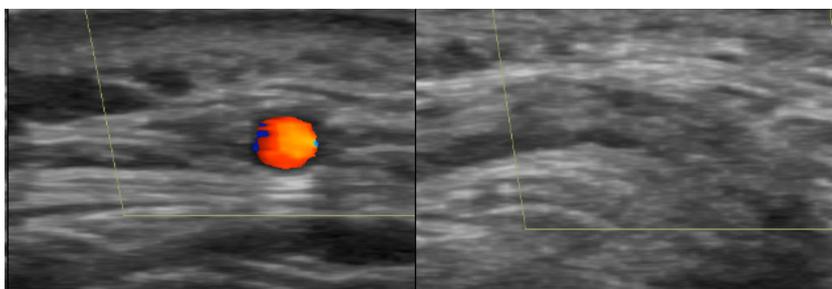
Slope Sign

The 'slope' sign in the axillary arteries is an ultrasound sign which may help you differentiate between a thickened IMT in Large Vessel Vasculitis (LVV) and atherosclerosis as it delineates the concentric thickening of vasculitis (10).

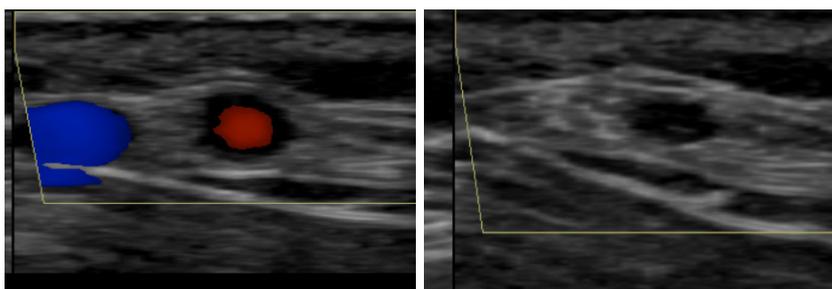


Compression Sign

- 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.
- The two layers of the thickened arterial wall remain visible upon compression; the hypoechoic, vasculitic vessel wall thickening contrasts against the mid-echoic to hyper-echoic surrounding soft tissues.
- The compression sign reflects the double wall IMT measurement.



Normal Compression: the vessel disappears completely on compression – compression sign negative



Abnormal Compression: the thickened vessel wall remains visible on compression, giving the appearance of a non-compressible halo described as “compression sign positive” (11).

8. Confounding Factors and Pitfalls

- Remember that Ultrasound is not 100% and a negative scan does not necessarily mean there is no GCA but it is useful information for experienced clinicians within a defined GCA pathway
- Make note if your patient has been on steroids prior to your scan, as this can lead to false negative results.
- Hair - ensure the hair is parted to fill the gaps with “stripes of gel” where required
- Males have thicker arterial walls - be wary of calling this thickened intima
- Atherosclerosis - asymmetric v symmetric wall irregularity. Hyperechoic changes within the intima are not necessarily related to GCA and should be described as such in your report any intimal defect/asymmetry should be described in the report– however clearly state if there is no Halo present. The SVT protocol includes Pulsed Doppler evaluation to assess for stenosis <https://www.svtgbi.org.uk/professional-issues/>
- Densely calcified vessels - this can make them impossible to compress, but is rare
- Tortuous vessels - ensure you have the necessary skills to follow the vessels accurately. The key is to ensure scanning in cross section and longitudinal to ensure all vessels are assessed along their entire length
- Inadequate colour Doppler settings can skew results. Ensure optimal colour Doppler settings to achieve optimal vessel filling without causing blooming artefact or under filling of the vessel axillary artery.
- Failure to use Zoom when measuring or measuring too close to arterial bifurcation
- Very narrow/stenotic lumen - can be easily compressed in error, mimicking vessel occlusion, however, if the vessel is diseased and accidentally compressed the ‘halo’ will be present which should alert sonographer to further assess

9. Reporting

Depending on departmental protocols, a descriptive report or more objective proforma may be used. Some examples are shown below.

Ultrasound Scan for Giant Cell Arteritis (GCA)			
Patient Name:		Date of Referral:	
NHS number:		Consultant:	
DOB:		Name of Sonographer:	
Or CRIS Label			
Contact Telephone Number		Scan Date:	
IMAGING INFORMATION			
<i>Complete all fields that apply:</i>			
Steroids given	<input type="checkbox"/> Yes <input type="checkbox"/> No	Date Steroids started	
Is this a repeat scan?	<input type="checkbox"/> Yes <input type="checkbox"/> No		
Date of Onset of Symptoms		Biopsy taken? Please state site	
RIGHT	Imaged	Halo Present	Compression Positive
Common Temporal	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N
Parietal	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N
Frontal	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N
Axilla	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	IMT thickness.....mms
LEFT	Imaged	Halo Present	Compression Positive
Common Temporal	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N
Parietal	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N
Frontal	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N
Axilla	<input type="checkbox"/> Y <input type="checkbox"/> N	<input type="checkbox"/> Y <input type="checkbox"/> N	IMT thickness.....mms
Ultrasound Result: GCA confirmed? <input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Equivocal If yes, please state site			
Comments: List any problems with the patient scan and reason for any images not provided or an equivocal result.			

Sample Normal Report 1

Doppler Ultrasound Axillary and Temporal arteries for GCA

Both superficial temporal and axillary arteries were examined.

Ultrasound temporal arteries: the common temporal, parietal and frontal branches were visualised bilaterally.

Right STA - The right common temporal, frontal and parietal branches of the superficial temporal artery are patent and are compression sign negative, with no halo visible - No evidence of GCA in these three branches.

Left STA - The left common temporal, frontal and parietal branches of the superficial temporal artery are patent and are compression sign negative, with no halo visible - No evidence of GCA in these three branches.

Ultrasound axillary arteries: both axillary arteries show normal colour filling and a normal IMT, with no halo seen.

Right Axillary IMT = mm.

Left Axillary IMT = mm.

Conclusion: No evidence of GCA was demonstrated.

Sample Abnormal Report 1

Sample Normal Report 2

Doppler Ultrasound Axillary and Temporal Arteries for GCA

The superficial temporal arteries and its frontal and parietal branches are widely patent and compressible. The axillary arteries are widely patent, with normal filling on colour Doppler. No intimal thickening evident

Conclusion: No ultrasound evidence of GCA

DOPPLER U/S BOTH AXILLARY AND TEMPORAL ARTERIES FOR GCA

Both axillary arteries show reduced colour filling of the lumen and an abnormal IMT, with positive halo sign seen. Right Axillary IMT = mm. Left Axillary IMT = mm.

Both superficial temporal arteries were examined.

Both common temporal, parietal and frontal branches were visualised.

Right STA - The right common temporal, frontal and parietal branches of the superficial temporal artery are all compression sign positive, with a positive halo sign visible - consistent with GCA in all three branches.

Left STA - The left common temporal artery shows a positive halo and compression sign, consistent with GCA. The left frontal and parietal branches of the superficial temporal artery are patent and are compression sign negative, with no halo visible - no evidence of GCA in these two branches.

Conclusion: Ultrasound appearances are consistent with temporal and axillary GCA

Sample Abnormal Report 2

Doppler ultrasound axillary and temporal arteries for GCA

There is abnormal, thickening of the intima giving a positive Halo sign (name vessel branch and location). At this point the vessel is visibly narrowed and not fully compressible. Ultrasound appearances are highly suspicious for GCA.

There is intimal thickening of the right common branch up to 0.6mm. Flow is visibly turbulent through a visible narrowing in the vessel which does not compress fully. There is intimal thickening to a lesser degree in the left parietal and frontal branches up to 0.3mm. No intimal thickening seen elsewhere in the temporal arteries or axillary arteries.

Conclusion - Ultrasound appearances are highly suspicious of GCA.

Sample Equivocal Report

DOPPLER U/S BOTH AXILLARY AND TEMPORAL ARTERIES FOR GCA

Both axillary arteries show normal colour filling and a normal IMT, with no halo seen.

Right Axillary IMT = mm.

Left Axillary IMT = mm.

Both superficial temporal arteries were examined.

Both common temporal, parietal and frontal branches were visualised.

Right STA - The right common temporal, frontal and parietal branches of the superficial temporal artery are patent and are compression sign negative, with no halo visible

- No evidence of GCA in these three branches.

Left STA - The left common temporal artery shows a positive compression sign, suggestive of GCA, but no obvious or definite halo sign was seen. This could also be due to atherosclerotic disease.

The left frontal and parietal branches of the superficial temporal artery are patent and are compression sign negative, with no halo visible - no evidence of GCA in these two branches.

Conclusion: Indeterminate scan - the appearances could suggest GCA, but all the positive features of GCA are not present. However, the patient has already had 9 days of high dose steroids which could affect the longevity of the halo sign and this is the patient's symptomatic side.

Cannot exclude GCA and TAB should be considered, dependent on the patients symptoms and their response to steroids. Equivocal GCA Scan

10. Other Learning Resources

 <p>BMUS THE BRITISH MEDICAL ULTRASOUND SOCIETY</p>	https://www.bmus.org
 <p>British Society for Rheumatology</p>	https://www.rheumatology.org.uk
 <p>VERSUS ARTHRITIS</p>	https://www.versusarthritis.org
 <p>eular fighting rheumatic & musculoskeletal diseases together</p>	https://www.eular.org
 <p>THE SOCIETY FOR VASCULAR TECHNOLOGY OF GREAT BRITAIN AND IRELAND</p>	https://www.svtgbi.org.uk
 <p>VASCULITIS UK</p>	https://www.vasculitis.org.uk

11. References

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Acknowledgements

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