Temporal arteries Doppler Ultrasound for the diagnosis of Giant Cell Arteritis

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Learning objectives

To describe the epidemiology, typical presentation and diagnostic criteria of Giant Cell Arteritis (GCA), as well as the current treatment.

To explain temporal arteries Doppler ultrasound technique, and enlist its advantages and limitations in the diagnosis of GCA, as comparing to the current diagnostic gold standard, temporal artery biopsy.

To describe and illustrate normal and pathological findings.

Background

- Epidemiology:

GCA is the most common vasculitis diagnosed in western countries, with an incidence rounding 20 per 100,000 older than 50 years.

It affects women two to six times more than man, and it is almost exclusive to patients over the age of 50 years. Its incidence increases with age.

- Etiology:

It is a systemic vasculitis, of unknown etiology, that affects large and medium sized vessels, such as the aorta and its branches, and extra cranial branches of the carotid artery, with typical involvement of the temporal arteries. It can also affect small vessels such as ophthalmic and ciliary arteries.

Histologically it is characterized by transmural inflammation of the vascular wall (with patchy infiltration by lymphocytes, macrophages and giant cells), and skip lesions (segments of the artery affected, interspaced with normal wall).

- Associations:

GCA is closely associated with polymyalgia rheumatica, which is present in half of the patients.

- Clinical presentation
GCA presents typically with systemic, cranial, ophthalmic and neurologic symptoms: asthenia, anorexia, weight loss, new onset headache, jaw claudication, temporal artery tenderness and erythema, visual disturbances, low grade fever

- Complications

The most dreaded complication is irreversible visual loss caused by involvement of the ophthalmic artery. GCA is one of the main causes of acquired blindness. Other possible complications include ischemic stroke, aortic aneurism and coronary ischemia.

- Diagnosis

A correct diagnostic is vital, not only because of the serious complications of the disease, but also because of the risks of the treatment itself.

The American College of Rheumatology developed diagnostic criteria for giant cell arteritis:

1. Age at disease onset >=50 years
2. New headache
3. Temporal artery abnormality (tenderness or decreased pulsation)
4. Elevated erythrocyte sedimentation rate
5. Abnormal artery biopsy

The diagnosis is made when three or more criteria are present.

Temporal artery biopsy is the diagnostic gold-standard, with a sensitivity of 15- 40% and specificity of 100%, however a negative biopsy does not exclude GCA. It’s possible complications include hematoma, wound infection, facial paresis and skin necrosis.

- Treatment

Prompt treatment with corticosteroids is necessary to prevent serious complications such as permanent visual loss. The mean duration of the treatment is roughly 2 years.

Findings and procedure details
- **Technique:**

The patient should be placed in a supine position, with modest elevation of the head. The head should be facing the opposite side of the one being approached.

A high frequency linear transducer should be used, at least 9MHZ, but ideally 10-15MHz.

Both arteries should be assessed in longitudinal and transverse plans, starting at the common superficial temporal arteries anteriorly to the ear and continuing distally to the parietal and temporal branches.

The arteries should be evaluated on B-mode imaging, color flow and spectral/ pulsed wave Doppler.

Peak systolic velocities and end diastolic velocities should be obtained in all aspect of the arteries, namely anteriorly to the ear, before the bifurcation, and at the frontal and parietal branches.

Care should be taken in color Doppler so that color covers the artery lumen but does not "bleed" over into the outer walls obscuring wall inflammation, and also to maximize beam steering.

A correct angle of 45-60° with respect to the blood flow should be used.

The pulse repetition frequency should be optimized, approximately at 2.5KHz.

- **Findings:**

Normal superficial temporal arteries show thin walls, and no lumen narrowing or areas suggesting stenosis.

There are three Doppler ultrasound findings suggestive of GCA: the presence on a hypoechoic halo surrounding the artery, known as the halo sign, stenosis and occlusion.

1. The halo sign refers to a hypoechoic, circumferential wall thickening, ranging from 0.3mm to 2mm, which traduces inflammatory edema/swelling of the vascular wall.
2. Stenosis, characterized by a narrowing of the vessel lumen, is defined as a segmental increase in peak systolic velocity more than twice of that recorded in pre-stenotic segment.

3. Occlusion of the vessel, in which case no flow is detected after proper adjustment of color Doppler and pulsed wave Doppler parameters.

- Advantages:

Doppler ultrasound is a noninvasive, well-tolerated procedure, widely available, and relatively inexpensive.

In contrast to temporal artery biopsy, it is a somewhat quick procedure, and allows a prompt result, which is crucial for diagnosis and therapy initiation.

With Doppler ultrasound all the length of the artery can be evaluated, as opposed to biopsy, and this is of major importance given the segmental nature of the disease.

Sensitivity and specificity vary among studies, with sensitivity values between 68 and 95%, and specificity from 91 to 100%, emphasizing the high diagnostic accuracy of this procedure.

- Limitations:

The technique depends on the experience and skill of the examiner, hence it should be carried out by a sonographer with experience in Doppler ultrasound, and who has investigated at least 30 people without GCA.

One other limitation is the inability of the procedure to differentiate GCA from other vasculitis that can affect temporal arteries, such as Wegener's granulomatosis, Churg Strauss syndrome and microscopic polyangiitis.

Differentiation from atherosclerosis may be difficult as it can be responsible for elevations in peak systolic velocities and occlusion, however atherosclerotic plaques are usually more echogenic.

The sensitivity of this procedure, as well as the sensitivity of the biopsy, lowers after the beginning of treatment, as the findings described above disappear generally after 14 to 22 days of treatment, although they can persist for months. To avoid this limitation the procedure should be performed during the first two weeks of treatment.
Fig. 1: Normal temporal artery, with thin wall and no lumen narrowing.

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**Fig. 2:** Hypoechoic halo surrounding superficial temporal artery, longitudinal plane.

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**Fig. 3:** Hypoechoic halo surrounding superficial temporal artery, transverse plane.

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**Fig. 4:** Superficial temporal artery with hypoechoic halo turbulent flow, and areas of lumen narrowing suggesting stenosis.

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Fig. 6: Same patient as Fig. 7. Prestenotic segment, with a peak systolic velocity of 52.2 centimeters per second.

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Fig. 7: Same patient as Fig. 6, demonstrating stenosis: an increase in peak systolic velocity from 52 to 109 centimeters per second, twice the value obtained in the prestenotic segment.

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Fig. 5: Lumen narrowing suggesting stenosis, which was considered suspect of GCA in the clinical scenario. However, biopsy showed atherosclerosis.

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Conclusion

Diagnosis of GCA is based on a combination of clinical, laboratorial, imaging and histological findings, with temporal artery biopsy being executed in most patients, and currently considered gold-standard diagnostic tool, despite the invasiveness and possible complications associated.

Doppler ultrasound of the temporal arteries is a noninvasive, well-tolerated, relatively inexpensive procedure with sensitivity and specificity comparable to temporal artery histology.

Based on the available evidence, it should precede biopsy in every patient with suspicion of GCA.

In the near future, Doppler ultrasonography, when positive, may replace temporal artery biopsy in the diagnosis of this vasculitis in patients with typical symptoms, reserving biopsy for those with atypical symptoms or negative Doppler ultrasound.

For the present time, while the use of Doppler ultrasound is not established as a standard, it can be used to guide biopsy, mapping areas of affected artery, in order to lower false diagnostic results due to skip lesions.

Some studies also suggest the use of Doppler sonography as a tool in the follow up evaluation, demonstrating resolution of the pathological findings with appropriate treatment.

Personal information

References


G. Ciancio, M. Bruschi et al, "Ultrasonography in Diagnosis and Follow-Up of Temporal Arteritis: An Update", Challenges in Rheumatology, 2011;129-143.


