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CLINICAL UP-DATE

Giant cell arteritis: Diagnosis and treatment☆

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Abstract Giant cell arteritis is the most common primary systemic vasculitis in adults. The condition is granulomatous arteritis of large and medium vessels, which occurs almost exclusively in patients aged 50 years or more. This article reviews the diagnosis and treatment of the disease.

PALABRAS CLAVE
Arteritis de células gigantes;
Vasculitis;
Biopsia de arteria temporal

Arteritis de células gigantes: diagnóstico y tratamiento

Resumen La arteritis de células gigantes es la vasculitis sistémica primaria más frecuente en el adulto. Es una arteritis granulomatosa de grandes y medianos vasos, que ocurre casi exclusivamente en mayores de 50 años. Este artículo revisa el diagnóstico y el tratamiento de esta entidad.

Case report

A 76-year-old woman consulted for holocranial headache and mandibular claudication of 3 weeks of evolution. The examination of the temporal arteries was normal. Laboratory tests resulted in the following data: hemoglobin, 10.9 mg/dL; erythrocyte sedimentation rate (ESR), 78 mm/h; and C-reactive protein (CRP), 6.8 mg/dL (normal up to 1 mg/dL). A 5-mm long biopsy of the right temporal artery showed no relevant disorders. What is the best diagnostic–therapeutic approach?

Background

Giant cell arteritis (GCA), also known as temporal arteritis, is the most common primary systemic vasculitis in adults.[1,2] GCA is a granulomatous arteritis of large and medium

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vessels, which affects the territory of the temporal artery (among others) and occurs almost exclusively in patients older than 50 years.[1,2]

Diagnosis

The clinical suspicion of GCA should be established by the presence of manifestations such as cranial arteritis, extracranial arteritis, neurological and vestibular manifestations and systemic symptoms (Table 1).[1–6] Approximately 40–60% of patients with GCA have polymyalgia rheumatica, characterized by pain and stiffness in the shoulder and pelvic girdles.[1–5]

The most consistently altered laboratory parameter is the increase in acute phase reactants, such as ESR and CRP.[1–5,7–9] However, we occasionally find patients with a normal ESR and CRP.[8,9] Statins and nonsteroidal anti-inflammatory drugs seem to reduce ESR but not CRP.[10] Anemia of chronic disease, thrombocytosis, leukocytosis and increased hepatic enzyme levels are relatively common.[1–5,7] Antiferritin antibodies are present in more than 90% of patients,[11] but its diagnostic importance is unclear.

GCA should be confirmed through biopsy, typically from the temporal artery.[1,2] It is a simple technique that has almost no complications, although there have been reported cases of facial nerve injury.[1] The biopsy shows vasculitis with a predominantly mononuclear infiltrate and/or granulomas, at times with giant multinucleated cells.[1,2] The vascular inflammation is usually transmural, extending to the tunica media. However, there have been reports of patterns of inflammation limited to the small vessels surrounding the adventitia, to the adventitial vasa vasorum or to the entire tunica adventitia with no involvement of the tunica media.[12] A negative temporal artery biopsy does not rule out a diagnosis of GCA.[1,12,13] The vasculitic involvement is patchy; a sample of at least 10–20 mm in length is therefore recommended.[13–15] Samples measuring less than 5 mm in length seldom yield positive results.[15] For patients with a high clinical suspicion of GCA and a negative initial biopsy, repeating the biopsy in the contralateral temporal artery might be reasonable.[16] The European League Against Rheumatism (EULAR) recommends always performing a temporal artery biopsy at least 10 mm in length and considers contralateral artery biopsy useful in selected cases.[17] The British Society for Rheumatology (BSR) similarly recommends performing temporal artery biopsies at least 10 mm in length (ideally 20 mm or more) and considers contralateral biopsy in cases with a negative initial biopsy of a suboptimal length.[18] GCA can probably be ruled out in patients with a low clinical suspicion and a negative biopsy.[16] Treatment with corticosteroids, even for several weeks, does not reduce the diagnostic yield of temporal artery biopsies.[19,20] Temporal artery biopsy is not recommended for patients with polymyalgia rheumatica, with no clinical suspicion of GCA.[1] In 1990, the American College of Rheumatology established criteria for classifying GCA: age older than 50 years, recent headache, abnormal temporal arteries in the examination, ESR >50 mm/h and positive temporal artery biopsy.[21] These criteria are only qualifiers, and their diagnostic utility is questionable.[21,22]

Duplex ultrasonography of the temporal arteries can be useful in the diagnosis of GCA.[23,24] The most consistently reported finding is the halo sign, a concentric hypoechoic thickening of the arterial wall.[23,24] A number of authors have also suggested that a compatible duplex ultrasonography finding could be sufficient to establish the diagnosis of GCA, without the need for a biopsy.[24] It should be noted that the halo sign is infrequently observed in cases with no transmural inflammation.[25] Duplex ultrasonography can also demonstrate extracranial arterial involvement in some patients.[26] For example, data suggestive of vertebral arteritis helps establish the diagnosis of GCA in patients who present with a stroke in the vertebrobasilar territory.[27] High-resolution magnetic resonance imaging can show transmural thickening and contrast enhancement in the superficial cranial arteries,[28] as well as aortic arch involvement and that of its branches.[29] Positron emission tomography can show increased uptake in the large vessels.[30] Both imaging tests can be useful for patients with atypical presentations and for those with a high suspicion of GCA and negative temporal artery biopsy.[31]

For patients in whom GCA is suspected clinically and all available diagnostic tests have been exhausted and other diagnoses have been reasonably ruled out, treatment with corticosteroids may be initiated. If there is a clinical and analytical response, we will establish the presumptive diagnosis of GCA, remaining alert during follow-up to the possibility of an alternative diagnosis.[16] Fig. 1 outlines a proposal for a personal diagnostic algorithm for patients with suspected GCA.

Table 1 Clinical manifestations of giant cell arteritis.

Cranial arteritis	Headache
	Scalp pain
	Mandibular and lingual claudication
	Tender and thickened temporal arteries
	Transient or permanent visual disorders
	Diplopia
Extracranial arteritis	Aortitis
	Aortic aneurysm and dissection
	Aortic valve insufficiency
	Upper extremity arterial ischemia
	Lower extremity arterial ischemia
Neurological and vestibular manifestations	Ischemic stroke
	Dementia
	Polyneuropathy, mononeuropathy, plexopathy
	Audio-vestibular dysfunction
Systemic manifestations	Fever-febricula
	General syndrome
	Polymyalgia rheumatica

Source: Waldman et al.,[1] Calvo-Romero,[2] González-Gay et al.,[3] Armona et al.,[4] Calvo Romero et al.[5] and Bongartz et al.[6]