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Diagnosis and management of giant cell arteritis

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Submitted: 26 September 2011; **final acceptance:** 1 December 2011.

©British Journal of General Practice 2012; 62: 329–330.

DOI: 10.3399/bjgp12X649313

INTRODUCTION

Giant cell arteritis (GCA) is the commonest form of large-vessel vasculitis and affects branches of the external carotid artery but also the ciliary and retinal arteries. The symptoms are caused by local ischaemia due to endovascular damage and cytokine-mediated systemic illness. There is considerable overlap with polymyalgia rheumatica (PMR): 16–21% of patients with PMR have GCA on temporal artery biopsy, and symptoms of PMR are present in 40–60% of patients with GCA.[1] GCA occurs in 2.2 per 10 000 patient-years in the UK.[2] A full-time GP may expect to see one new case every 1–2 years. It is virtually unknown in people aged under 50 years. Early recognition is critical to prevent visual loss, that otherwise occurs in up to 20% of cases.[3] Once high-dose corticosteroids are started, visual loss is extremely rare.

Guidelines for the diagnosis and management of GCA, have recently been published by the British Society of Rheumatologists and British Health Professionals in Rheumatology.[4]

DIAGNOSIS

A 2002 systematic review analysed the presenting clinical features in a mixture of studies, with a total of 1435 cases of giant cell arteritis.[5] The mean duration of symptoms at diagnosis was 3.5 months.[5] The results in Table 1 demonstrate the somewhat protean manifestations of this condition.

The sensitivity of individual clinical features was relatively low: 24% of cases had no headache at all, and only 52% had a temporal headache. The diagnosis is easily missed when systemic symptoms (such as low-grade fever or weight loss), ischaemic symptoms (jaw claudication or transient visual symptoms), or polymyalgic symptoms (proximal myalgia or morning stiffness) predominate over the well-known hallmark of temporal headache. Unfortunately, there is some evidence that this subgroup (without headache as the dominant symptom) may be at increased risk of visual loss. A recent audit of 65 patients with GCA showed that 44 had had unrecognised visual disturbance, visual loss, or stroke in the mean of 35 days between onset of symptoms and diagnosis (range 2–336 days).[6] Eleven of these patients presented without headache or scalp tenderness, and 10 of these had visual loss.

Only 4% of patients with GCA have a completely 'normal' erythrocyte sedimentation rate (ESR) but nearly one-fifth have an ESR >50 mm/hour.

There is some evidence that GCA may be underdiagnosed. A 1971 Swedish study examined 1097 consecutive autopsies, with temporal artery examination carried out in each of them. Sixteen cases of undiagnosed GCA were identified (1.5% of the study population). Retrospective analysis of the case notes documented typical features of undiagnosed GCA in nine.[7]

Localities need to have a clear pathway for suspected GCA because GPs are often uncertain whether to refer to rheumatologists, ophthalmologists, or vascular surgeons.

Temporal artery ultrasound may become more used in diagnosis. A meta-analysis of studies of temporal artery ultrasound against a gold standard of temporal artery biopsy found a sensitivity of 69% and a specificity of 82%.[8]

BIOPSY

Urgent referral for specialist assessment and temporal artery biopsy is suggested for all patients with suspected GCA, although this should not delay initiation of immediate corticosteroid treatment. The biopsy can retain the characteristic giant cell histology for 2–6 weeks after initiation of treatment but should ideally be done within 2 weeks. The biopsy may be negative in 13% of true

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Provenance

Freely submitted; not externally peer reviewed.

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Table 1. Manifestations of giant cell arteritis

Clinical feature	Percentage of biopsy-proven cases with the feature (sensitivity)
Temporal headache	52
Any headache	76
Scalp tenderness	31
Jaw claudication	34
Any visual symptom	37
Unilateral visual loss	24
Diplopia	9
Myalgia	39
Previous diagnosis of PMR	34
Weight loss	43
Fever	42
Absent temporal pulse	45
Any abnormality on palpation of the temporal artery (absent, prominent, beaded)	65
ESR 'normal'	4
ESR >50 mm/h	83

ESR = erythrocyte sedimentation rate. PMR = polymyalgia rheumatica.

cases (possibly because of 'skip lesions').[9] If the clinical features are typical, the patients should, nevertheless, be treated.

TREATMENT

High-dose glucocorticosteroid therapy should be initiated immediately the diagnosis is suspected. There are few risks in starting treatment erroneously (the treatment can always be stopped) and delayed treatment can result in sudden visual loss. In the absence of ischaemic symptoms (jaw claudication or visual symptoms), it is reasonable to start on 40 mg prednisolone daily orally. If the patient has jaw claudication, the risk of visual loss is high and 60 mg prednisolone should be used. If the patient already has visual symptoms of any sort, then immediate admission for 3 days of intravenous methylprednisolone is necessary to preserve vision.

The initial dose of oral prednisolone is maintained until symptoms have resolved and inflammatory mediators have normalised. The dose can then be reduced by 10 mg at 2-week intervals until the patient is taking 20 mg daily, and then reduced by 2.5 mg steps each 2 weeks to 10 mg. Thereafter, a reduction of 1 mg per month every 4–8 weeks is recommended, as with PMR. Most patients have stopped treatment by 2 years. Review with measured inflammatory markers is initially weekly, tapering to monthly, and then 3-monthly.

Low-dose aspirin should be considered in those patients without contraindications and bisphosphonates with calcium and vitamin D supplementation are recommended for all patients on long-term corticosteroids.

Relapse is usually, but not always, associated with a rise in inflammatory markers. Rarely, patients may develop a more widespread vasculitis of the aortic arch and its branches. Upper-limb claudication, absent pulses, or widening of the mediastinum on a chest X-ray should prompt urgent specialist evaluation.

PATIENT EDUCATION AND SELF-MANAGEMENT

Patients should receive written information on GCA (such as the Arthritis Research Campaign booklet on GCA), together with instructions about seeking urgent review in the event of any return of symptoms. Support can be obtained from the local Polymalgia Rheumatica and Giant Cell Arteritis UK (PMRGCAUK) patient support group (http://www.pmrgcauk.com/).

CONCLUSION

GCA is the commonest form of vasculitis. A GP will encounter a new case roughly once every 1–2 years. Early recognition and treatment with high-dose corticosteroids is crucial to preventing the visual loss that occurs in 20% of patients. There is evidence that the risk of visual loss is higher in patients with jaw claudication and in patients who do not have the typical temporal headache. The starting dose for prednisolone is 40 mg to 60 mg. Specialist referral is advised and temporal artery biopsy should ideally occur within 2 weeks.