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Diagnosis of giant cell arteritis: when should we biopsy the temporal artery?

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Abstract

Giant cell arteritis (GCA) can be diagnosed histopathologically by biopsy of the temporal artery, and clinically using the 5-point score of the 1990 American College of Rheumatology (ACR) classification. We aimed to find out whether some patients are referred for biopsy unnecessarily. We audited all referrals (n = 100) made to the Department of Oral and Maxillofacial Surgery over 34 months, and used the ACR classification to find out whether patients had had a clinical diagnosis of GCA at referral (ACR score: 3 or more). We then compared them with the result of the biopsy. Of the 100 referred, 98 had a biopsy, and of them, 15 were diagnosed with GCA (2 results were not included). Thirteen of the 15 had already been diagnosed clinically (based on the ACR classification) at referral. Our results gave an ACR specificity of 96% (95% CI: 85% to 99%) but only 20% sensitivity (95% CI: 11% to 32%). There was a linear correlation of high ACR scores with histopathological confirmation. Biopsy is most beneficial when there is a degree of diagnostic uncertainty (ACR: 1 or 2), an atypical presentation, or when steroids may be relatively contraindicated. On the basis of our study, we designed a new referral form for biopsy based on the ACR criteria.
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Keywords: Giant Cell Arteritis; Temporal Cell Arteritis; Temporal artery biopsy

Introduction

Giant cell arteritis (GCA), also commonly known as temporal arteritis, is a chronic, granulomatous vasculitis that affects medium to large vessels, predominantly branches of the common carotid artery. It is the most common form of vasculitis, with a reported UK incidence of 2.2 cases/10 000 patient years.[1]

GCA occurs almost exclusively in patients over 50 years of age, usually in those around 70, and is 3 times more prevalent in women than in men.[1] Symptoms include sudden-onset headache (which is often unilateral and localised to the temporal region), claudication of the jaw, and visual disturbances, most notably transient monocular visual loss (amaurosis fugax), which can lead to blindness.[1] The danger of acute blindness resulting from retinal ischaemia[1] makes the disease a true medical emergency that warrants prompt diagnosis and treatment with high-dose corticosteroids, usually prednisolone 40 – 60 mg daily.[1]

Temporal artery biopsy has long been considered the gold standard investigation to help confirm a clinical diagnosis of temporal or giant cell arteritis. The characteristic histopathological appearance of granulomatous inflammation with infiltration of giant cells in the arterial walls, notably within the

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tunica media, is so specific to the disease (100%) that some consider analysis of a biopsy specimen to be the best way to confirm the clinical diagnosis[2] and to guide the management of the patient. The principle disadvantage is the possibility of a false negative result.[3–5] Biopsy can also cause complications, and although the risk of serious morbidity is low, the facial nerve can be injured, and postoperative problems such as haemorrhage, scarring, pain, and infection, can occur.[6,7]

The clinical classification of GCA, which was formalised in 1990 by the American College of Rheumatology (ACR score) in its guidelines on the differentiation of giant cell arteritis from other forms of vasculitis,[8] was originally developed for research purposes, but claims of a specificity of 91.2% and sensitivity of 93.5% led to its clinical use.[8] The scoring criteria include 5 equally weighted factors: age over 50 years; new-onset, localised headache; tenderness around, or decreased pulse in, the temporal artery; erythrocyte sedimentation rate (ESR) of at least 50 mm in the first hour; and an abnormal artery specimen characterised by mononuclear infiltration or granulomatous inflammation. GCA, as opposed to any other vasculitis, is highly probable in patients with 3 or more of the above.

In our hospital, all biopsies of the temporal artery are done under local anaesthetic. The procedure takes 30 minutes and costs £800. Specimens are usually harvested unilaterally depending on the clinical symptoms, as bilateral specimens give an additional diagnostic yield of only 1% - 5%.[9] In line with recommendations, we try to obtain specimens that are at least 20 mm long.[10] Requests for biopsy are often urgent and samples are harvested at the earliest opportunity. However, although we were sent a relatively high volume of requests, most specimens showed no evidence of GCA, and in many instances, the result did not seem to alter the subsequent management of the patient. We therefore aimed to examine the results from our existing referral system and, if necessary, streamline the indications for biopsy. On the basis of previous small studies[11–13] we hypothesised that histopathological confirmation would be likely in patients who presented with at least 3 of the ACR criteria, and that the ACR scores could ultimately be used to reduce the number referred for biopsy.

Method

We studied data from all patients referred to the Department of Oral and Maxillofacial Surgery for biopsy of the temporal artery over 34 months (Oct 2008 - July 2011). Identification was both retrospective (from a pre-existing departmental database) and prospective (from incoming referral letters). We used the ACR classification to establish whether a patient had been diagnosed clinically at referral (ACR score of 3 or more), and compared it to the result of the biopsy.

We obtained patients' details, their presenting symptoms and histopathological or blood results from the referral letter or case notes, and the hospital's database. The dates of referral and biopsy, ACR score at presentation, biopsy result, length of the specimen, and steroid dose, were recorded.[7]

Sensitivity was calculated using true positives or true positives plus false negatives. Specificity was calculated using true negatives or false positives plus true negatives. A histopathological diagnosis was considered the gold standard. Confidence intervals (CI) were calculated at 95%. The correlation co-efficient (r) value was also calculated, and values between 0.4 and 0.6 were considered to have a reasonable positive linear relation.

Results

A total of 100 patients were referred for biopsy (65 by hospital physicians, 20 by general practitioners, and 15 were not known). There were 31 men and 69 women (ratio 1:2), median age at referral 72 years (range 43-93). Two refused the procedure, and of the 98 specimens, one was venous rather than arterial, and another could not be traced.

A total of 34 (34%) had complained of some form of visual disturbance at presentation. The mean length of the specimens was 30 mm (range 5-60), which is more than the recommended minimum of 20 mm.[10] The shortest one showed features of GCA.

In 15 of the 96 biopsies (16%) GCA was confirmed, and in 81 (84%) it was not. Of the 98 patients referred, more than half (n = 52, 53%) met the ACR criteria preoperatively for a diagnosis of GCA (Table 1). Of those confirmed cases, 13 of 15 had already met the ACR criteria for a diagnosis before operation. According to our results, the preoperative scores had a specificity of 96% (95% CI: 86% to 99%) but a sensitivity of only 20% (95% CI: 11% to 32%).

We found that the likelihood of histopathologically confirmed disease increased the higher the score, giving reasonable linear correlation when r = 0.6 (Table 2). Because disease was confirmed in few patients with a score of 1 or 2, but in a large proportion of those with a score of 3 or more, our results suggest that a score of 3 is an appropriate cut-off point for the clinical prediction of disease that will be confirmed histopathologically.

Before biopsy, 95% (95/100) of patients had been started on steroids. Of the 96 biopsy results available, data on the interval between referral and operation were available for 76

Table 1
American College of Rheumatology (ACR) scores at referral and number of cases of giant cell arteritis confirmed on temporal artery biopsy. Data are number (%).

ACR score	No. of patients (n = 98*)	No. of cases confirmed histopathologically
3-5	52 (53)*	13/51 (26)
1-2	46 (48)**	2/45 (4)

* 2/98 biopsy results were not included: *1 report was untraceable, **1 was a venous sample.

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