BMJ

BMJ 2011;342:d3019 doi: 10.1136/bmj.d3019

PRACTICE

EASILY MISSED?

Giant cell arteritis

Nada Hassan *specialist registrar*[1], Bhaskar Dasgupta *consultant rheumatologist*[1], Kevin Barraclough *general practitioner*[2]

[1]Rheumatology, Southend University Hospital, Westcliff-on-Sea SS0 0RY, UK; [2]Greenbank, Painswick GL6 6TY, UK

This is one of a series of occasional articles highlighting conditions that may be more common than many doctors realise or may be missed at first presentation. The series advisers are Anthony Harnden, university lecturer in general practice, Department of Primary Health Care, University of Oxford, and Richard Lehman, general practitioner, Banbury. To suggest a topic for this series, please email us at easilymissed@bmj.com.

Giant cell arteritis affects large and medium sized arteries, often 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: 16-21% of patients with polymyalgia rheumatica have giant cell arteritis on temporal artery biopsy, and symptoms of polymyalgia rheumatica are present in 40-60% of patients with giant cell arteritis.[1]

Why is giant cell arteritis missed?

A systematic review analysed the presenting clinical features in a mixture of studies with a total of 1435 cases of giant cell arteritis.[3] The sensitivity of individual clinical features was relatively low (table), reflecting the diverse presentation of this condition: 24% of cases had no headache at all, and only 52% had a temporal headache. Giant cell arteritis can be 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. The mean duration of symptoms in the 1435 patients at diagnosis was 3.5 months. A 1971 Swedish study examined 1097 consecutive autopsies with temporal artery examination carried out in each of them. Sixteen cases of undiagnosed giant cell arteritis were identified. Retrospective analysis of the case notes documented typical features of undiagnosed giant cell arteritis in 9.[4] A recent audit of 65 patients with giant cell arteritis 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 of 2 to 336 days).[5] Eleven of these patients presented without headache or scalp tenderness and 10 of these had visual loss.

Why does this matter?

Acute blindness occurs in up to 20% of patients with giant cell arteritis.[4] Delay in recognition may explain the high incidence of irreversible loss of vision, which is preventable with early diagnosis and treatment.[4] Jaw or tongue claudication occurs in a minority of cases but heralds a high rsik of impending ischaemic complications.[6]

How is giant cell arteritis diagnosed?

The typical presentation of giant cell arteritis is with temporal headache of recent onset, myalgia, or systemic malaise or fever, and the mean age of onset is 70. Erythrocyte sedimentation rate or C reactive protein is typically raised.[2] The case presented illustrates that “typical” features may be absent or subtle.

A meta-analysis looked at studies examining the value of individual clinical features in predicting positive results of temporal artery biopsy in patients with suspected giant cell arteritis.[3] No clinical features had a high negative likelihood ratio, because no clinical feature (even headache) was reliably present in all cases. Several symptoms were moderately predictive of a positive biopsy result (likelihood ratio >2):

- Jaw claudication (present in 34% of cases); claudicant pain comes on gradually during chewing, whereas temperomandibular pain or dental pain is immediate
- Diplopia (present 8% of cases)
- Any abnormality on palpation of the temporal artery—absent, beaded, tender, or enlarged (present in 65% of cases).

Other useful predictive features (likelihood ratio >1.5) were

- Temporal headache
- Scalp tenderness
- Erythrocyte sedimentation rate >100 mm/h

Correspondence to: K Barraclough k.barraclough@btinternet.com

Case scenario

A previously fit and well 72 year old man presented to his general practitioner after several months of malaise and weight loss. When asked about any pain, he pointed to his left scalp as being painful. Examination was normal except that his left temporal artery was not palpable. Suspecting giant cell arteritis, the GP started 40 mg prednisolone and requested an erythrocyte sedimentation rate, which was 86 mm/h. The symptoms largely disappeared in 48 hours. She also referred the patient for a temporal artery biopsy, which showed giant cell arteritis.

How common is it?

- Giant cell arteritis occurs in 2.2 per 10 000 patient years in the United Kingdom[2]
- A full time general practitioner may expect to see one new case every 1-2 years
- It is virtually unknown in people aged under 50

- Anaemia.

The limitation of these studies is that giant cell arteritis was already highly likely, as all patients in the studied population had biopsies. In practice, the condition should be suspected in anyone over the age of 50 with headache, scalp tenderness, transient visual symptoms, or unexplained facial pain. Examination may show no abnormalities, but palpation of the temporal artery is often abnormal. Only 4% of patients have a completely "normal" erythrocyte sedimentation rate (half the age of the patient, plus 5 for women[7]); 83% have a rate above 50 mm/h.Once giant cell arteritis is suspected, refer patients urgently for temporal artery biopsy, which needs to be done within two weeks of starting steroids. The true sensitivity of temporal biopsy is not known, but one model estimates a sensitivity of 87%.[8]

How is giant cell arteritis managed?

Most treatment recommendations are based on experts' opinion, as randomised trials would be unethical.[7] Once the diagnosis is suspected, treat with high dose corticosteroid immediately. Give 40 mg prednisolone daily unless the patient has ischaemic symptoms (jaw or tongue claudication, or visual symptoms). With claudication symptoms, give 60 mg prednisolone daily; if the patient has visual symptoms, admit for treatment with intravenous methylprednisolone.Once symptoms and abnormal test results resolve, the dose can be reduced in 10 mg steps each two weeks to 20 mg, then in 2.5 mg steps. Most patients have stopped taking steroids by two years.[9]

Contributors: KB conceived the idea. NH and BD wrote the first draft; KB revised it. All authors approved the article before publication. KB is guarantor.

Funding: No additional funding.

Competing interests: All authors have completed the Unified Competing Interest form at www.icmje.org/coi_disclosure.pdf and declare: no support from any organisation for the submitted work; no financial relationships with any organisations that might have an interest in the submitted work in the previous three years. KB and BD were authors of the 2010 British Society of Rheumatologists guidelines on the management of giant cell arteritis.

Provenance and peer review: Not commissioned; externally peer reviewed.

Patient consent not required (patient anonymised, dead, or hypothetical).

1 Salvarani C, Cantini F, Hunder GG. Polymyalgia rheumatica and giant-cell arteritis. *Lancet* 2008;372:234-45.
2 Smeeth L, Cook C, Hall AJ. Incidence of diagnosed polymyalgia rheumatica and temporal arteritis in the United Kingdom, 1990 to 2001. *Ann Rheum Dis* 2006;65:1093-8.
3 Smetana GW, Shmerling RH. Does this patient have temporal arteritis? *JAMA* 2002;287:92-101.
4 Ostberg G. Temporal arteritis in a large necropsy series. *Ann Rheum Dis* 1971;30:224-35.
5 Ezeonyeji AN, Borg FA, Dasgupta B. Delays in recognition and management of giant cell arteritis: results from a retrospective audit. *Clin Rheumatol* 2011;30:259-62.
6 Salvarni C, Cimino L, Macchioni P, Consonni D, Cantinni F, Bajocchi G, et al. Risk factors for visual loss in an Italian population-based cohort of patients with giant cell arteritis. *Arthritis Rheum* 2005;53:293-7.
7 Miller A, Green M, Robinson D. Simple rule for calculating normal erythrocyte sedimentation rate. *BMJ* 1983;286:266.
8 Niederkohr RD, Levin LA. A Bayesian analysis of the true sensitivity of a temporal artery biopsy. *Invest Opthalmol Vis Sci* 2007;48:675-80.
9 Dasgupta B, Borg FA, Hassan N, Alexander L, Barraclough K, Bourke B, et al. BSR and BHPR guidelines for the management of giant cell arteritis. *Rheumatology* 2010;keq039a.

Accepted: 28 January 2011

Cite this as: *BMJ* 2011;342:d3019

Learning points

- Giant cell arteritis is a medical emergency, as irreversible loss of vision can occur in 20% of cases without prompt treatment with steroids
- "Typical" features such as headache and scalp tenderness may be absent, and this subgroup has a high risk of visual loss
- Jaw or tongue claudication occurs in a minority of cases and heralds impending ischaemic complications
- Erythrocyte sedimentation rate and C-reactive protein are normal in only 4% of cases but may be just mildly raised in a fifth of cases
- If giant cell arteritis is suspected, immediate treatment with high dose corticosteroids is indicated, and temporal biopsy should be arranged within two weeks of starting treatment

Table

Table 1| Sensitivity of clinical features in predicting giant cell arteritis3

Clinical feature	% of biopsy proven cases (sensitivity)
Headache:	
Any headache	76
Temporal headache	52
Scalp tenderness	31
Jaw claudication	34
Visual symptoms:	
Any visual symptom	37
Unilateral visual loss	24
Diplopia	9
Myalgia	39
Previous diagnosis of polymyalgia rheumatica	34
Weight loss	43
Fever	42
Temporal artery:	
Pulse absent	45
Any abnormality on palpation of temporal artery (absent, prominent, beaded)	65
Erythrocyte sedimentation rate:	
"Normal"	4
>50 mm/h	83