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CASE REPORT

Temporal arteritis with erythrocyte sedimentation rate <50 mm/h: a clinical reminder

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Abstract: Temporal arteritis, also known as giant cell arteritis (GCA), is a systemic vasculitis that predominantly involves the temporal arteries. It is a medical emergency and should be treated promptly as it can lead to permanent loss of vision. It is very commonly associated with a raised erythrocyte sedimentation rate (ESR), usually >50 mm/h, one of the essential criteria defined by the American College of Rheumatology classification of GCA. Here, we describe the case of a 73-year-old male presenting with a 2-day history of a sudden onset of a severe left-sided headache, which had the signs and symptoms consistent with GCA but he had an ESR of only 27 mm/h. The patient was urgently treated with prednisolone 60 mg per day, and his symptoms dramatically improved within 24 hours of therapy. Temporal artery biopsy results were consistent with an inflammatory response, and withdrawal of treatment led to a relapse of the symptoms. The patient was slowly tapered off the high steroid dose and is now currently managed on a low steroid dose. We should keep a high index of suspicion for GCA in patients presenting with clinical symptoms of GCA even though the ESR is <50 mm/h as stated in the criteria for GCA diagnosis.

Keywords: temporal arteritis, giant cell arteritis, prednisolone, erythrocyte sedimentation rate

Case history

A 73-year-old male presented to the Medical Assessment Unit with sudden onset of a 2-day history of severe left-sided headache. He described the headache as dull and throbbing, predominantly localized in the frontal and temporal areas on the left side radiating down to the left side of the neck. On further examination, mild temporal artery beading could be palpated. In addition, the patient had mild nausea and slight photophobia since the headache began. There were no focal neurological deficits or meningism and no preceding aura. On physical examination, he was afebrile and vital signs were stable; however, he had an exquisitely tender left temporal artery. Fundoscopic examination did not reveal any abnormalities. A complete vascular examination was performed, which did not reveal any upper extremity pulse loss or subclavian, carotid, and axillary bruits. There was no asymmetry of pulses and blood pressure in the extremities. Computed tomography of the head ruled out any space-occupying lesions or hemorrhagic lesions. Lumbar puncture was performed to screen for infection and xanthochromia (both negative). Past medical history included well-controlled chronic obstructive airway disease with good exercise tolerance for independent daily activities.

A full blood count showed a normal white cell count of 9.2×10^9/L with only positive findings of a raised erythrocyte sedimentation rate (ESR) of 27 mm/h (normal range: 1–15 mm/h) and a slightly raised C-reactive protein of 10 mg/L (normal range: <5 mg/L). Chest radiograph and urinalysis did not reveal any abnormalities. The patient was urgently prescribed 60 mg (0.75–1 mg/kg) of prednisolone as per hospital protocol, and during treatment, he had regular follow-up to monitor for complications of high-dose steroid

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therapy. He had symptomatic relief within 24 hours of initiation of steroid therapy. According to local hospital protocol, the patient underwent an urgent temporal artery biopsy (TAB) within 4 weeks after the steroid therapy was initiated, and it suggested signs of postinflammatory changes consistent with vasculitic changes and reduplication of internal elastic lamina modified by steroid therapy. These signs were consistent with an inflammatory response, which is responding effectively to steroid therapy. The patient was prescribed bisphosphonates and calcium and vitamin D supplements for bone protection. A proton pump inhibitor was prescribed to provide gastric protection. The patient was followed up in rheumatology and ophthalmology departments as per hospital protocol. He was followed up initially at 0, 1, 3, and 6 weeks, and then at 3, 6, 9, and 12 months in the first year.[1] Additional visits were encouraged if signs of relapse or adverse events were observed. Rapid relapse of symptoms was observed after withdrawing glucocorticosteroids, which excluded vascular and neurological causes for patient symptoms. During treatment, the patient had regular glucose monitoring, blood pressure recordings, visual field testing, and assessment for side effects of corticosteroid therapy. The patient made a complete recovery, and symptoms are now under control with low-dose (1 mg) prednisolone for long-term maintenance treatment. Ethical approval was sought from the Research and Development Department at Wirral University Teaching Hospital. Verbal informed consent was taken from the patient to publish the case study.

Discussion

The prevalence of giant cell arteritis (GCA) in the general population is <1%.[2] GCA is a large-vessel vasculitis, and it usually affects individuals >50 years old.[2] The disease is unlikely to occur in individuals <50 years of age.[2] GCA is surprisingly a frequently occurring condition with a prevalence estimated to be 1 in 500 in individuals >50 years[3] and incidence of 2.2 per 10,000 patient-years in the UK.[4] The mean age for GCA occurrence is 72 years,[2] as GCA is a type of systemic vasculitis whose symptomatology is quite extensive. Thus, the presentation of the disease can be quite variable and diverse. The symptoms could include constitutional symptoms such as tiredness, fever, and weight loss. Fever is usually of low grade; however, high-grade fever of >39°C has been reported in 15% of patients.[5] Other clinical features include abrupt-onset headache, which is usually unilateral affecting the temporal region; however, diffuse and bilateral headaches have been reported;[1,4] scalp pain or localized scalp tenderness; jaw claudication; nonproductive cough;[6] and visual symptoms such as amaurosis fugax, blurring of vision, or even blindness in severe cases.

Other associated conditions include polymyalgia rheumatica and, in chronic cases, aortic involvement leading to aneurysms and aortic dissection.[2] Aortic disease features are usually a consequence of long-term, low-grade vasculitis and typically not present at the time of diagnosis. Less common complications may include dysarthria,[7] throat pain and tongue infarction,[8] mononeuritis multiplex,[9] sensorineural hearing loss,[10] and, even rarely, mesenteric ischemia.[11] According to the American College of Rheumatology (ACR), diagnosis of GCA should be considered in all patients aged >50 years who have at least three of the five criteria findings (Table 1). The gold standard test to confirm the diagnosis is TAB, which should be performed in all patients suspected of GCA; however, consideration should be given to the urgency of initiation of medical treatment and waiting for biopsy should not delay initiation of high-dose steroid treatment in a suspected case of GCA.[1] The positivity of the biopsy results can be variable and is dependent on numerous factors. These include sampling error due to skip lesions, attaining too small a sample (<2 cm), and different phenotypic disease not associated with cranial arteritis in which case the TAB will be negative even if repeated.[12] Such patients may have GCA but only involving large vessels, including subclavian artery (arm claudication), carotid artery, and the aorta (aneurysms and dissection),[12] but without cranial vessel involvement. These patients will not require TAB; however, they may need systemic imaging to assess isolated large-vessel GCA. Early TAB should be performed preferably within 1–2 weeks of initiating glucocorticoids; however, reports suggest that results may remain positive for 2–6 weeks following initiation of glucocorticosteroids.[13,14]

Duplex ultrasonography secures a promising role in the future for diagnosis of GCA, particularly large-vessel disease; however, it does not currently replace TAB as the first-line investigation as recommended by the British Society of Rheumatology guidelines.[1] It is user dependent and requires a high level of expertise, which are yet not currently

Table 1 Criteria used to diagnose temporal arteritis

American College of Rheumatology criteria for temporal arteritis
Age ≥50 years
New onset or new type of localized pain in the head
Elevated ESR >50 mm/h (by Westergren method)
Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
Biopsy specimen showing vasculitis characterized by mononuclear infiltration or granulomatous inflammation, usually with multinucleated giant cells

Note: The patient is said to have temporal arteritis if at least three out of five criteria are present. Data from Hunder et al.[15]
Abbreviation: ESR, erythrocyte sedimentation rate.

widespread in UK. Furthermore, it does not have the added prognostic value of histology. In case of large-vessel GCA, ultrasound is still a sensitive technique, especially for upper limb vasculitides.

Historically, ESR has been considered one of the most useful markers to predict the likelihood of having GCA. A normal ESR makes GCA unlikely; however, ESR does not rule it out.[2] A meta-analysis of 114 studies showed that normal ESR values indicate much less likelihood of a positive diagnosis of GCA (negative likelihood ratio (LR) for abnormal ESR, 0.2; 95% confidence interval [CI], 0.08–0.51), but physical findings such as temporal artery tenderness indicated a higher likelihood of GCA (positive LR, 2.6; 95% CI, 1.9–3.7).[2] This meta-analysis revealed that a high level of ESR was a less important indicator in ruling out GCA as the underlying cause for the patient's symptoms, but positive physical findings, characteristic of GCA, were more likely to be a strong indicator of a positive diagnosis of GCA. Our case reported to have positive clinical findings such as temporal artery tenderness with a mildly raised ESR; however, ESR was not raised to fulfill the ACR criteria for GCA.

Treatment includes immediate initiation of high-dose glucocorticosteroids, recommended upon suspicion of GCA.[1] Visual loss is an early complication of the disease; however, once established, it rarely improves, and hence, the emphasis is on early treatment. Prednisolone 40–60 mg daily is usually the recommended dose for 4 weeks. The total duration of high-dose prednisolone is usually governed clinically by the resolution of symptoms and improvement in laboratory test abnormalities.[16,17] The dose is then reduced by 10 mg every 2 weeks up to 20 mg, then by 2.5 mg every 2–4 weeks up to 10 mg, and then by 1 mg every 1–2 months, provided there are no further relapses.[18] These patients should also be coprescribed bone protection (weekly bisphosphonates and calcium/vitamin D supplementation).[19] Gastrointestinal protection with proton pump inhibitors should also be used. Considering that these patients are on a high dose of glucocorticosteroids, they require close monitoring of adverse events, which can be possible through shared care with primary care physicians.

Monitoring and follow-up

At each visit, the following investigations should be performed:[1]

- full blood count, ESR, C-reactive protein, urea and electrolytes, and glucose (to look for steroid-induced diabetes);
- every 2 years, chest radiograph to monitor for aortic aneurysm (more detailed investigations such as MRI and echocardiography may also be appropriate);
- as patients are on long-term steroid therapy, bone mineral density scans may be required.

Follow-up schedules can vary according to local hospital protocols. British Society of Rheumatology recommend close monitoring at weeks 0, 1, 3, and 6 and then at months 3, 6, 9, and 12 in the first year.[1] Unscheduled visits are advised in the event of relapse.

Treatment of relapse follows a similar regimen:[1]

- Headache: patients should be restarted on the previous higher prednisolone dosage.
- Headache and jaw claudication: treatment should be started with 60 mg prednisolone.
- Visual symptoms: treat with either 60 mg prednisolone or intravenous methylprednisolone.
- Large-vessel GCA: further investigation such as positron emission tomography and MRI imaging is recommended and consider treatments using systemic vasculitis protocols.

Predictors of neuro-ophthalmic (cranial) complications

Recent study evaluating risks of cranial ischemic event in GCA patients revealed that patients with low systemic inflammatory response (odds ratio [OR] =0.30, 95% CI, 0.08–1.08), hypertension (OR =7.77, 95% CI, 0.83–72.76), and a past history of ischemic heart disease (OR =8.65, 95% CI, 0.92–80.95) are associated with a high risk of developing severe cranial ischemic events.[20] Low-dose aspirin has also been shown to decrease cranial complications.[1] In resistant or recurrent GCA, immunosuppressive agents such as methotrexate may be used as adjuvant therapy to allow reduction in glucocorticosteroid use.

A meta-analysis identified different relationships between clinical features and TAB positivity.[2] A positive TAB has been associated with cranial complications, including cerebrovascular strokes.[2] Clinical features that increase the likelihood of having a positive TAB include jaw claudication: shows a high positive LR 4.2 (2.8–6.2), diplopia: LR 3.4 (1.3–8.6), temporal artery beading: LR 4.6 (1.1–18.4), and temporal artery tenderness: LR 2.6 (1.9–3.7).

Features that have been shown to have less likelihood of a positive TAB include the absence of temporal artery abnormality (beading, tenderness): negative LR 0.53 (0.38–0.75) and a normal value of ESR: negative LR 0.2 (0.08–0.51).[2]

Conclusion

The diagnosis of temporal arteritis requires a high index of suspicion as it may manifest in a variety of clinical features. However, a mildly elevated ESR in the presence of positive

characteristic clinical features is increasingly suggestive of GCA and should still trigger initiation of treatment for GCA. Acute medical professionals need to be aware of the criteria of GCA but, in addition, need to be aware of atypical GCA presentations such as low ESR, arm claudication, dysarthria, and phenotypic cases not involving cranial arteries.

Key learning points

- Consider diagnosis of temporal arteritis in patients with new onset or new type of localized headache having age ≥50 years.
- Consider diagnosis of temporal arteritis even in patients with ESR <50 mmHg/h but who meet the criteria of temporal arteritis clinically.
- Prompt diagnosis and urgent treatment should be initiated by the acute medical doctors for temporal arteritis.

Acknowledgment

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Disclosure

The authors report no conflicts of interest in this work.

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