Giant Cell Arteritis Affecting the Tongue: A Case Report and Review of the Literature

Abstract: Giant cell arteritis (GCA) is a systemic vasculitis with symptoms that could cause a patient to present to a general dental practitioner. A case of GCA that presented as headache, jaw claudication, unilateral visual loss and tongue ulceration leading to necrosis is reported and the literature reviewed, with an emphasis on dentally relevant aspects. It is vital that GCA is not overlooked in patients over the age of 50 with unexplained dental pain, tissue necrosis or jaw pain which may be misdiagnosed as a temporomandibular joint disorder. Early diagnosis and prompt treatment is the key to preventing visual loss. Early referral in such cases would be warranted.

Clinical Relevance: Dental clinicians may play a part in the early diagnosis of GCA by having a high index of suspicion for its symptoms in patients, so that devastating ischaemic consequences, such as irreversible visual loss, can be prevented.

Dent Update 2013; 40: 669–677

Giant cell arteritis (GCA) is the most common vasculitis of adults in the western world.1 It is a systemic inflammatory vasculitis that affects medium- to large-sized arteries. Arterial wall inflammation leads to arterial occlusion and tissue ischaemia, which cause the clinical manifestations of vasculitis.2 Early diagnosis and prompt treatment is the key to preventing devastating ischaemic consequences, such as permanent visual loss.3

Some manifestations of GCA may lead to the patient seeking the advice of a dentist. These include unexplained dental pain, oral mucosal necrosis and jaw pain, which may be misdiagnosed as temporomandibular dysfunction (TMJ). Jones and Hazleman4 described three cases that initially presented to the dentist but were not recognized as GCA. Hence, steroid treatment was delayed and the patient remained at risk of blindness.

A case is reported where GCA presented as a headache, jaw pain, necrosis of the tongue and unilateral blindness and the literature relating to GCA is reviewed, particularly in relation to oral and maxillofacial symptoms.

Case report

A 79-year-old female was seen in an emergency ophthalmology clinic presenting with sudden complete visual loss in the left eye in addition to a history of a generalized ongoing headache of two weeks’ duration. The pain affected the back of the head, ear and jaws, which worsened on mastication. The patient also reported a history of pain when moving her tongue, which recently developed a large necrotic ulcer. Previously, the right-hand side of her face had swollen.

Medically she had hypertension, osteoporosis and osteoarthritis and was taking tramadol, bendroflumethiazide, lercanidipine, perindopril, paracetamol, calcichew, alendronic acid and omeprazole. The patient had never smoked and had minimal alcohol intake.

On extra-oral examination, left and right temporal arteries were tender and firm on palpation. An examination by ophthalmology revealed a pale optic disc in the left eye. Intra-oral examination revealed a large necrotic ulcer on the right dorsal, lateral border and ventral surface of the anterior tongue.

Haematological investigations revealed a markedly elevated erythrocyte sedimentation rate (ESR) of 68mm/hr and C-reactive protein (CRP) of 150mg/l, reflecting the increased level of systemic inflammation. A full blood count revealed thrombocytosis 535x10^9/l (increased platelets, normal range 150–400x10^9/l), neutrophilia 10.79x10^9/l (increased neutrophils, normal range 2.00–8.00x10^9/l) and an erythrocyte macrocytosis MCV 100fl (increased red blood cell size, normal range 80.0–98.0fl).

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A diagnosis of GCA with associated anterior ischaemic optic neuropathy and tongue necrosis was made. Bilateral symptoms indicate the systemic nature of the condition.

The patient was started on oral prednisolone 60 mg daily. A rheumatology opinion confirmed the diagnosis of GCA, with the patient showing no other signs or symptoms to suggest any other connective tissue or vasculitic disorder. Following admission, the patient was pulsed with 500 mg methylprednisolone IV for three days in an attempt to reduce the underlying inflammation further. Following four days of high dose corticosteroid therapy, the patient’s ESR and CRP were gradually decreasing, but the tongue was deteriorating to the extent that eating was impossible. An urgent referral to oral medicine was arranged where the patient was seen by a consultant. Examination revealed a grossly necrotic tongue, with much of the right anterior tongue affected (Figure 1). Debridement of the necrotic tissue was undertaken with a scalpel (Figure 2). Topical application of 0.1% chlorhexidine gluconate mouthwash to the ulcer with a sponge was advised. Benzydamine hydrochloride spray (Difflam, Meda Pharmaceuticals, UK) was prescribed for symptomatic relief.

The patient was kept under outpatient review with rheumatology, ophthalmology and oral medicine. ESR and CRP continued to decrease and the oral prednisolone dose was gradually decreased to 10 mg daily over six months with no further exacerbations of GCA. Visual acuity in the right eye was 6/9. In the left eye only hand movements and perception of light were noted. The tongue healed well over the next 2 months, with only a small notch defect in the right lateral border, which was attributed to scar tissue. A small area of depapillation on the dorsal surface was present after six months (Figure 3 (a–c) – healing sequence).

This case demonstrates the importance of considering GCA in cases where unusual intra-oral ulceration is seen.

**Discussion**

**Definitions**

Giant cell arteritis is also known as temporal arteritis, cranial arteritis and Horton’s disease. The term GCA is commonly used to describe the multi-nucleate giant cells that are visible histologically in the walls of affected vessels. It is often referred to as temporal arteritis because of its propensity to involve the temporal arteries. The most susceptible sites for this disease are the medium and large extra-cranial branches of the carotid arteries. Involvement can extend to the aorta and its primary and secondary branches.

**Historical perspective**

Historically, the earliest alleged recorded observations of GCA were in the 10th Century. It was initially described in the English speaking literature in 1890, when a patient noted streaks on his head, so painful that it prevented him wearing his hat. These streaks were inflamed temporal arteries. The first histopathological evidence of granulomatous arteritis was by Horton in 1932, hence the eponym of Horton’s disease.

**Epidemiology**

The overall incidence ranges from 1.25 to 29.1 per 100,000 in individuals over the age of 50 years. Incidence increases with age, with median age of onset being 75 years. Women are affected two to six times more commonly than men. GCA is most frequent among people of Scandinavian and Northern European descent. It is infrequently found in Asians and Afro-Caribbeans. This, along with reports of familial clustering and concordance with monozygotic twins, suggests an inherited component to the disease.

**Pathogenesis overview**

The immunological events that lead to the granulomatous inflammation in the vessels are not fully understood.
Two different processes underlie the clinical manifestations of GCA. The trigger for these processes has only been hypothesized. The first is a systemic inflammatory reaction resulting from overactivation of the innate acute phase response, a non-antigen driven, non-adaptive defence mechanism to stress and injury. Manifestations related to this systemic response are non-specific markers for inflammation, including fever, myalgias, anorexia, weight loss and night sweats. The second process is a maladaptive antigen specific immune response directed to arterial walls and is responsible for the focal ischaemic complications of GCA.

Clinical features

Natural history

This is best revealed in the outcome of cases before the advent of steroid treatment. These showed that GCA was episodic in nature, lasting from a few months to three years or more, with exacerbations and remissions. Spontaneous recovery occurs even without corticosteroid therapy. This highlights the importance of retrospective history-taking rather than looking for symptoms at the time of presentation. Remission does not rule out later GCA complications.

Systemic manifestations

These include fever, malaise, proximal myopathy (ache when raising arms or getting out of a low chair), weight loss and arthralgia. One-third of patients with GCA concomitantly suffer with polymyalgia rheumatica (PMR) at presentation. PMR is characterized by myalgias (muscle pain) in the neck, shoulders and pelvic girdle.

Ophthalmological manifestations

Visual loss is the most common ophthalmological complication of GCA. Visual loss can be transient or permanent and results from insufficient perfusion of the optic nerve, retina or choroid. Transient monocular blindness (amaurosis fugax) has been shown to precede permanent visual loss in more than half of untreated cases by an average of 8.5 days. Thus, transient visual loss should still be considered to be an ophthalmological emergency. Permanent visual loss associated with GCA is most commonly due to anterior ischaemic optic neuropathy, caused by the inflammatory occlusion of the short posterior ciliary arteries.

Oral and maxillofacial manifestations

Headache and craniofacial pain

Pain occurs either in the area of arteritis or is due to distal ischaemia from an occluded artery.

Headache occurs in up to 90% of patients and can occur in any location, but patients are sometimes able to localize to temporal and occipital regions. Unilateral headache is rare. Scalp tenderness can manifest as pain on brushing or washing hair, putting a hat on or placing one’s head on a pillow. These symptoms are more indicative of tissue ischaemia.

Jaw claudication is an intense cramp-like, like pain on chewing or talking that decreases at rest, and is reported in 30–48% of patients. This may be so severe that it limits mouth-opening and may be misdiagnosed as TMJ dysfunction. Abnormal temporal arteries are present in only around 50% of cases and present as tenderness on palpation, loss of pulse and as an indurated or nodular artery. Other manifestations can include dental pain, facial nerve palsy, chin numbness, dysphagia, dysarthria and as a submandibular mass. Other areas affected by tissue necrosis include the scalp, lips and nasal septum. Facial swelling most frequently presents as fullness of the tissues overlying the zygoma, maxilla and around the orbit.

Lingual manifestations

Tongue necrosis has been noted as a rare but dramatic manifestation of GCA. Missen showed that on autopsy of five patients that died of GCA, nine out of ten lingual arteries showed subclinical ischaemia in 1959. Subsequently, it has been frequently noted, gaining increasing recognition as being a manifestation of GCA. Sixty-two case reports in the remaining literature. Eight cases in the English language have presented initially as tongue necrosis with a delay in diagnosis leading to blindness.

Tongue necrosis carries significant morbidity, with decreased mobility leading to slurred speech and eating problems. Morbidity following necrosis can be minimized if microcirculation is rapidly improved by the initiation of steroid therapy. Necrotic tissue should be debrided. Reports of auto-amputation have been seen, notably when one patient spat out the anterior two-thirds of their tongue. In cases where necrotic tissue has not been debrided, aspiration of tissue has followed and caused laryngeal obstruction and precipitated respiratory failure.

GCA is the most documented cause of necrosis of the tongue. Differential diagnoses for other causes of ischaemia or necrosis of the tongue are listed in Table 2.

General dental practitioners may be the first port of call for a patient suffering with signs such as tongue ulceration and necrosis. Damage to the retinal blood supply is likely to be advanced by the time of dental presentation.

Occult GCA

GCA can prove a diagnostic challenge when it doesn’t follow the classic sequence of disease progression.
Occult GCA has the potential to present as a dysfunction of an organ system in Table 3, with normal inflammatory markers.

**Investigations**

Erythrocyte sedimentation rate has been considered a hallmark test for diagnosis of GCA. An elevated ESR is a non-specific marker of inflammation. A low ESR, however, does not rule out GCA. C-reactive protein is an acute phase marker of inflammation, which has been shown to be more specific to GCA, with a sensitivity of 98–100%. Histologically, GCA is characterized by inflammation composed of CD4+ T lymphocytes, macrophages and giant cells affecting all layers of arteries that have an elastic lamina. Luminal stenosis or occlusion may be visualized.

**Diagnosis**

The American College of Rheumatology criteria for the diagnosis of GCA is shown in Table 4. The presence of any three of these five criteria permit a diagnosis of GCA to be made. These criteria have been shown to under-represent patients with ischaemic complications and those with occult GCA. CRP and imaging modalities were not included in the criteria. There has been a call for updating these criteria.

**Treatment**

Treatment is aimed at decreasing the inflammatory process in an attempt to prevent ischaemic complications. Corticosteroids remain the treatment modality of choice, not only due to their anti-inflammatory effects but also antioxidant and vasodilator effects. No randomized controlled trials have evaluated different steroid dosing regimens but the general consensus is that high-dose steroids started promptly is the best predictor of visual improvements. Generally, oral prednisolone at 1mg/kg/day is recommended. Methylprednisolone 250 mg IV can be given for the first 3–5 days, if necessary.

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**Table 1.** Signs and symptoms of developing GCA affecting the tongue.

<table>
<thead>
<tr>
<th>Sign/Symptom</th>
<th>Sign/Symptom</th>
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<tbody>
<tr>
<td>Claudication</td>
<td>Calciplaxis (vascular calciphylaxis)</td>
</tr>
<tr>
<td>Blanchning</td>
<td>Tuberculosis</td>
</tr>
<tr>
<td>Blotches</td>
<td>Radiotherapy</td>
</tr>
<tr>
<td>Loss of taste</td>
<td>Mid-tongue tumour</td>
</tr>
<tr>
<td>Loss of taste</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Loss of taste</td>
<td>Embolism</td>
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</tbody>
</table>

**Table 2.** Differential diagnoses for ischaemia or necrosis of the tongue.

<table>
<thead>
<tr>
<th>System</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiovascular</td>
<td>Aortic aneurysm, myocardial infarction</td>
</tr>
<tr>
<td>Ear, nose and throat</td>
<td>Hearing loss, tinnitus, vertigo and dizziness</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Bowel infarction from mesenteric artery occlusion</td>
</tr>
<tr>
<td>Neurological</td>
<td>Cerebrovascular ischaemic events, peripheral neuropathies</td>
</tr>
</tbody>
</table>

**Table 3.** Organ system manifestations of GCA.

<table>
<thead>
<tr>
<th>Number</th>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Age of onset greater than 50 years</td>
</tr>
<tr>
<td>2</td>
<td>Onset of new headache</td>
</tr>
<tr>
<td>3</td>
<td>Temporal artery abnormality (tenderness or reduced pulsation)</td>
</tr>
<tr>
<td>4</td>
<td>Elevated ESR &gt;50 mm/hr</td>
</tr>
<tr>
<td>5</td>
<td>Abnormal arterial biopsy</td>
</tr>
</tbody>
</table>

**Table 4.** American College of Rheumatology criteria for GCA diagnosis.
acute visual signs are seen, followed by a high oral dose for 4–6 weeks. Bisphosphonates are recommended to prevent glucocorticoid-induced osteoporosis and a proton pump inhibitor to reduce peptic ulceration should also be considered. Prednisolone can be reduced at 5–10 mg/month until 20–30 mg per day is reached. Thereafter, reduction by 2.5–5 mg a month can then be carried out until a maintenance dose of 10 mg daily is reached. If any recurrence of symptoms or a rise in ESR or CRP occurs the dose should be increased.5

Prognosis
Discontinuation of steroids is possible within the first few years whilst closely monitoring the ESR and CRP: but long-term therapy is often required and side-effects are therefore common. For the majority of patients suffering with loss of vision, it will not recover. Despite the risk of developing cardiovascular complications, no increased mortality with GCA has been shown.3

Conclusion
GCA is an important disease in relation to dentistry. Some symptoms may result in the patient consulting a general dental practitioner or hospital dental staff being requested to examine an inpatient. These can include unexplained dental pain, tissue necrosis or jaw pain, which may be misdiagnosed as a TMJ disorder. Dental clinicians may play a part in early diagnosis and should have a high index of suspicion for GCA in patients over 50 years of age so that devastating ischaemic consequences, such as irreversible visual loss, can be prevented. Early urgent referral would be warranted in suspected cases.

References