TEMPORAL ARTERITIS

An unusual presentation

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INTRODUCTION

Temporal arteritis (TA), also known as giant-cell arteritis, most commonly presents with headache and scalp tenderness affecting the lateral and temporal areas, tongue and jaw claudication, and altered visual disturbance. These symptoms relate to the anatomy and distribution of the arterial circulation, and if not recognised early, the condition can progress to cause blindness. The condition is not exclusive to the extra-cranial circulation, however, and can present with other, less common, manifestations leading to significant morbidity if not recognised and managed appropriately. The following case is an example of an atypical presentation of TA, demonstrating the diagnostic and management dilemmas that may arise.

CASE REPORT

An 82-year-old lady presented to the Primary Care Assessment Service at Westmorland General Hospital (WGH) with difficulty chewing, which she explained was due to her tongue going numb, which also made speech difficult. On further questioning it was found that she had also had a headache affecting the frontal area for approximately one week, but did not have any other focal neurological signs other than those affecting her tongue. She was otherwise well, only suffering from hypertension which was normally well controlled with medication. She had long-standing blindness in her right eye. TA was one of the differential diagnoses.

On admission to the ward, her observations were entirely normal except for a raised blood pressure (168/80). Examination was also unremarkable with no temporal artery tenderness. Her dysarthria persisted, with inability to put her tongue out. There was no obvious other abnormality of her tongue, and no other neurological deficit. Investigations revealed a raised white cell count (WCC) at 14.5, neutrophils 8.5, erythrocyte sedimentation rate (ESR) 61, C-reactive protein (CRP) 81, and a total cholesterol of 6. On the basis of the clinical picture and blood results a provisional diagnosis of TA was made and the patient was started on prednisolone 40mg. A temporal artery biopsy was booked. A computerised tomography brain scan was also requested to rule out stroke as a differential or coexisting diagnosis; the result was negative.

After several days her headache had improved significantly to almost complete resolution, but her tongue was no better and had developed white 'plaques' which, whilst affecting both sides of the tongue, were more prominent on the left.

The patient was referred to the maxillofacial surgeons for their advice regarding management as parts of her tongue were becoming necrotic. After ten days of steroid therapy the patient's ESR had fallen to 34, and her CRP was now 26. On reviewing the patient, the maxillofacial team felt that the appearance of the tongue was consistent with squamous cell carcinoma. A biopsy was taken and a magnetic resonance imaging (MRI) scan arranged for staging purposes. It was also felt that the patient should continue on the steroids.

The patient was seen at the regional unit as an outpatient within three days, where the diagnosis and treatment options of local surgical excision and radiotherapy were explained to her. Despite the tongue biopsy being negative, the opinion of the consultant was that this was a T4NxMx tumour (see table 1) and the patient was referred for dental assessment prior to dental extraction in preparation for oral radiotherapy. The medical team felt it best to defer this until the temporal artery biopsy was available.

Primary tumour
T0: No evidence of primary tumour
T1: Primary tumour <2 cm
T2: Primary tumour 2-4 cm
T3: Primary tumour >4 cm
T4: Invasion of tumour into adjacent structures (eg, muscle, skin, bone, nerves)

Nodal status
N0: No regional lymph node metastasis
N1: Metastasis to a single ipsilateral lymph node (<3 cm)
N2a: Metastasis to a single ipsilateral lymph node (3-6 cm)
N2b: Metastasis to multiple ipsilateral lymph nodes (<6 cm)
N2c: Metastasis to bilateral or contralateral lymph nodes (<6 cm)
N3: Metastasis to any lymph node (>6 cm)

Distant metastasis
Mx: Cannot be assessed
M0: No distant metastasis
M1: Distant metastasis

Staging
Stage 1: T1N0M0
Stage 2: T2N0M0
Stage 3: T3N0M0, T1N1M0, T2N1M0, T3N1M0
Stage 4: Any T4 lesion, any N2 or N3 lesion, any M1 lesion

Table 1 TNM staging for oral cancer

Very shortly after this, the temporal artery biopsy was reported as positive with evidence of inflammation consistent with TA found. The dose of prednisolone was increased to 50mg (two 25mg tablets) as the patient was struggling to swallow eight 5mg tablets, and a carotid ultrasound scan (USS) was requested to assess internal and external carotid arteries. The MRI scan was reported as '*artefact from metallic clips from the biopsy site. There is no evidence of tumour extension or cervical node enlargement.*'

A second tongue biopsy performed (due to concerns over the quality of the first one) reported that despite the presence of ulcerative and hyperplastic squamous mucosa, there was

no malignancy seen. By now the ESR was down to 18, and the CRP 8. The patient remained well in herself despite her painful tongue, which now started to show signs of recovery with the plaques beginning to slough off (see figure 1). Based on the result of the two biopsies and improvement in her tongue, radiotherapy was cancelled. The result of the carotid USS showed 55% stenosis of the left internal carotid artery only. The patient was discharged home on 35mg of prednisolone. Her tongue was almost fully healed and ESR was 16.

By October, three months after discharge, the patient was recovering well on 25mg prednisolone, ESR remained stable at 5, and further reduction in steroid dose planned. Other medications include aspirin 75mg, atenolol 25mg, bendrofluazide 2.5mg, lisinopril 10mg, simvastatin 40mg, alendronate 70mg weekly and adcal D3. Although she was started on dipyridamole, this has been discontinued as she has had issues swallowing it. Her tongue had recovered well (see figure 2), although only time will tell to what extent function returns to the affected area.

Figure 1 Appearance of tongue after three weeks on steroids. Photograph courtesy of Mr John Buchan – staff grade, maxillofacial surgeon, WGH

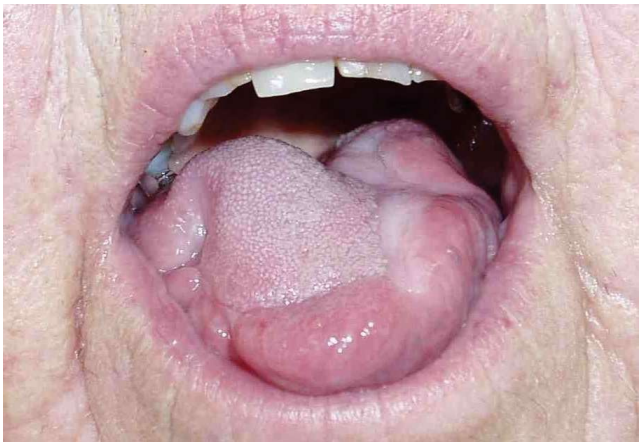

Figure 2 Appearance of tongue at three months

DISCUSSION

TA is a condition in which inappropriate activation of T cells leads to inflammation and hyperplasia of the walls of medium or large sized arteries,[1] thus causing ischaemia and necrosis of the anatomically affected area. As such it is a condition that all clinicians should be aware of as failure to recognise and manage it in a timely and appropriate manner can have disastrous consequences for the patient, most notably blindness secondary to ischaemic optic neuritis.[2]

Ordinarily, the condition has a predictable presentation of headache, associated with scalp tenderness in the distribution of the extra cranial arteries. Jaw or tongue claudication and visual disturbances are also common features.[3] A raised ESR gives the first clue, but the gold standard for diagnosis is a positive biopsy from an affected artery as soon as possible. Management is with longterm corticosteroid therapy, with a recommended starting dose of 40-60mg prednisolone.

Inflammed arteries are not exclusive to those of the scalp, however, and any medium to large artery may be involved leading to uncommon presentations that can be easily confused with other pathologies. Often these other presentations have their own distinguishing features such as those outlined in table 2.[4] This table demonstrates how in many cases, due to the inflammatory and occlusive nature of the condition, these features relate to the anatomical distribution of the affected arteries. Two lingual arteries, branches of the external carotids, supply the tongue. They originate adjacent to the tips of the greater horn of the hyoid bone, projecting forward towards the tongue's apex. They also supply the sublingual gland, gingiva and oral mucosa.[5]

Respiratory tract symptoms
Dry cough
Sore throat
Tongue pain
Choking sensation
Fever of unknown origin
Large artery involvement
Upper and lower extremity claudication
Thoracic or abdominal aneurysm
Peripheral nervous system features
Mononeuritis multiplex (especially of the brachial plexus)
Central nervous system features
Stroke
Transient ischaemic attacks
Dementia
Hallucinations
Syndrome of inappropriate antidiuretic hormone
Tumour-like lesions
Breast mass
Ovarian mass
Microangiopathic haemolytic anaemia

Table 2 Atypical manifestations of temporal arteritis[4]

Tongue necrosis itself is rare, with a number of causes including the use of ergotamine,[6,7] but the most common is said to be TA.[8] Not only is it rare, but its appearance can also be confused with other pathologies as demonstrated in the case presented here, thus adding to the difficulty in diagnosis, and potentially delaying treatment or instigating inappropriate treatment. Unfortunately the list of differentials is a significant one, each having significant implications if ignored in favour of another.

CONCLUSION

Early diagnosis and treatment of TA is imperative, but this can prove difficult if reliant on the result of biopsy. The longterm effects of corticosteroids are well recognised, but in the short term they are unlikely to be problematic and therefore should never be withheld in suspected cases of TA.

This case demonstrates the problem for a patient who has a rare condition where the differential diagnosis is also rare (the audit of head and neck cancer 2006-2007 in Lancaster showed one patient with tongue cancer that year)[9]. The rush to diagnose and treat patients with cancer led to this patient having unnecessary anxiety, particularly as she is the main carer for her husband, who has dementia. It also showed that getting a biopsy done and obtaining the results is quicker for suspected cancer than for non-cancer (such as a temporal artery biopsy) when both can be equally important. In this case although we were relatively confident our initial diagnosis was correct, we were concerned we had missed an oral cancer when the regional team were so sure, and found it difficult to give full reassurance to the patient that all would be well with steroid treatment.

It also demonstrates that caring for a patient on a medical ward needs continuity. If this patient had been handed on to a different team following the post-take ward round, the team's determination to continue with the treatment of TA may have been swayed with disastrous results, including the removal of the patient's teeth and the flare up of TA.

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