Unusual presentation of more common disease/injury

Temporal arteritis presenting with tongue necrosis

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Summary

Spontaneous glossodynia is uncommon and glossodynia progressing to necrosis is especially rare. Although the commonest cause of lingual necrosis is giant cell arteritis, only a few cases of a new diagnosis of giant cell arteritis, clinically presenting with isolated lingual necrosis, have been reported.

BACKGROUND

Giant cell arteritis (GCA) also known as temporal or cranial arteritis is a chronic, systemic vasculitis of large and medium-sized vessels, usually affecting the over-60 age group. With its preponderance to arteries of the head and neck, GCA often produces cranial and visual symptoms. The first reported case of lingual necrosis secondary to GCA was by Howard and Cremin¹ in 1959. Although there have been a number of cases reported since, GCA presenting with bilateral lingual necrosis as the primary clinical feature is rare.²⁻⁴ We present the unusual case of a 74-year-old woman who presented with glossodynia which progressed to acute lingual necrosis. Classical symptoms and signs of GCA were absent but the diagnosis was suspected clinically and later confirmed by temporal artery biopsy.

CASE PRESENTATION

A 74-year-old Caucasian woman presented as an emergency to a district general hospital complaining of an acutely painful, swollen and discoloured tongue. The patient had consulted her primary care physician earlier in the day complaining of mixed pain and numbness in her tongue, causing difficulties with eating. Numbness had developed on the left side of her tongue 3 days prior to this presentation, but resolved within 24 hours. Initial

clinical examination, however, was normal. In addition, she had also complained of sore throat in the preceding week, for which she had completed a 5 day course of erythromycin.

Her medical history included hypertension, hypercholesterolaemia and psoriasis. Her surgical history was of a hysterectomy and a tonsillectomy. She is a smoker of approximately six cigarettes per day.

Her symptoms deteriorated in the 24 h preceding presentation to the emergency department, such that speech and eating were impaired. Although she initially denied other symptoms, upon further specific questioning, she admitted to a vague history of mild headaches and jaw pain on chewing, in the 4 days preceding onset of tongue symptoms. There was no history of amaurosis fugax or diplopia. Further, there was no history of focal limb or facial weakness, or of limb claudication. She also denied fever, malaise or myalgia.

On examination in the emergency department, the tongue was noted to be swollen with slight blue discoloration (figure 1A). Within 12 h, the tongue became exquisitely tender and swollen with grey-white discoloration on the dorsal surface and a well-demarcated triangular blue/black area on the tip with two deep fissures. She was afebrile and there was no palpable lymphadenopathy. In addition, there was no temporal artery tenderness,

Figure 1 Initial progression of lingual necrosis despite medical therapy (A, B) followed by surgical resection of necrotic tissue (C) and subsequent clinical improvement (1D). (A) Appearance of tongue at initial presentation. (B) Day 8 of hospital admission: well-demarcated area of grey/white discoloration of tip of tongue with slough. (C) Day 8: surgical debridement of tongue tissue (intraoperative). (D) Appearance of tongue at outpatient follow-up 28 days after initial presentation.

prominence or nodularity. No focal cranial or peripheral neurology was found on formal examination. All peripheral pulses were palpable and there were no vascular bruits.

INVESTIGATIONS

Blood results on admission revealed a raised erythrocyte sedimentation rate (ESR) and C reactive protein at 103 mm/h and 37 mg/l, respectively. Full blood count revealed a neutrophil leucocytosis at $15.4 \times 10^9/l$ and a reactive thrombocytosis at $792 \times 10^9/l$. A tongue swab grew candida species. Therefore, she was commenced on a course of fluconazole and nystatin oral suspension. An MRI of her brain, neck and oral cavity showed no evidence of a brainstem or other intracranial vascular pathology. Nasal endoscopy excluded any other coexisting lesions or abnormalities in the nasopharynx. She was also assessed by ophthalmology and no significant ocular pathology was identified. Most importantly, there were no signs of anterior ischaemic optic neuropathy. She was negative for antinuclear antibody and antineutrophil cytoplasmic antibody.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included an underlying vasculitic process such as GCA, thromboembolic event, necrotising glossitis, hypersensitivity reaction and 'black hairy tongue'. Antihistamines and corticosteroids were administered in the emergency department, as hypersensitivity reaction was the working diagnosis at the time.

TREATMENT

The patient was reviewed shortly after admission by a rheumatologist and, suspecting an underlying vasculitic process, commenced prednisolone 40 mg once daily along with aspirin at an initial loading dose of 300 mg followed by a maintenance dose of 75 mg once daily. In addition, the patient received thromboprophylaxis with dalaprin 5000 units daily and used thromboembolic deterrent stockings (TEDS). In addition to regular analgesia, she required intravenous fluids as lingual pain limited oral fluid intake. Headaches improved after commencement of steroids; however, the clinical appearance of the tongue

became more necrotic with on-going severe pain and raised inflammatory markers (figure 1B).

Eight days after her presentation, her care was transferred to the maxillo-facial surgery team for debridement of necrotic tip of tongue under general anaesthetic, as well as a diagnostic temporal artery biopsy. Intraoperatively, necrotic tissue sloughed off easily but revealed underlying ischaemia. The tongue was resected until the bleeding fissure (figure 1C). Right and left lingual nerves were both identified. Sublingual ducts and mylohyoid muscle were not visualised. The defect was closed primarily and resected tongue tissue was sent for histology. Concurrently, a 2 cm biopsy specimen of the left temporal artery was obtained—the biopsy procedure was straightforward and uneventful.

OUTCOME AND FOLLOW-UP

In the tongue sample, histology confirmed non-specific ischaemic changes and areas of infarction with marked ulceration and distortion of normal tissue architecture, but no discernible histological features of an arteritis or vasculitis (figure 2A). However, the left temporal artery biopsy (figure 2B,C), performed on clinical suspicion of GCA, revealed extensive transmural infiltrate of lymphocytes and histiocytes with characteristic giant cells, extending into the adventitial tissues. A thrombus occluded the vessel lumen and there was evidence of focal disruption of the internal elastic lamina, a typical feature of temporal arteritis.

The final diagnosis was therefore of lingual necrosis secondary to GCA.

Postoperatively she was given prophylactic antibiotics and initially fed via a nasogastric tube, followed by slow build up to purée foods, solids and liquids. She received input from the dieticians, and speech and language therapists. Having made a good postoperative recovery, she was discharged after a 16-day hospital stay. She was discharged on aspirin 75 mg daily and a tapering course of prednisolone, together with omeprazole and alendronic acid for gastric and bone protection, respectively. Despite the partial loss of tongue tissue, she recovered well and attained near-normal speech and function at discharge. At a recent review in rheumatology outpatients, our patient remains well with complete resolution of her original

Figure 2 Histological appearance of tongue (A) and temporal artery biopsy (2B, C). (A) Resected tongue tissue showing non-specific ischaemic changes and areas of infarction with marked ulceration and distortion of normal tissue architecture. (B, C) Temporal artery biopsy showing extensive transmural infiltrate of lymphocytes and histiocytes (black frame) with characteristic giant cells (thick arrows), extending into the adventitial tissues. A thrombus occluded the vessel lumen (thin arrow).

symptoms (figure 1D). Her latest ESR and C reactive protein are 5 mm/h and <1 mg/l, respectively. She continues to taper prednisolone in 5 mg steps monthly.

DISCUSSION

We report a patient whose first presentation of previously undiagnosed GCA was with lingual ischaemia progressing to necrosis. Numbness, pain and necrosis of the tongue are all rare symptoms which necessitate consideration of a wide range of differential diagnoses by the attending clinician. Tongue numbness and pain without any clinical signs implies an endogenous, haematological or endocrinological abnormality and maybe the earliest feature suggestive of lingual ischaemia.⁵ Lingual necrosis in itself is an especially rare manifestation as the rich collateral supply of the tongue would be expected to provide a compensatory vascular supply.

GCA primarily presenting as lingual necrosis, without any other constitutional or focal temporal symptoms is especially rare. Alternative aetiology of lingual necrosis ranges from embolism, lingual carcinoma and regional radiotherapy to Wegener's granulomatosis and ergotamine-containing drugs.⁵⁻⁷ However, the commonest cause of lingual necrosis is reported to be GCA.

GCA is a systemic disease of unknown aetiology, characterised by granulomatous infiltration of the walls of medium and large-sized arteries. It is thought to be immune mediated with a genetic component as well. Genes associated with human leucocyte antigen DR4 haplotypes, female sex and northern European descent are believed to predispose to the development of GCA.⁸ The immune hypothesis is that microorganisms, ranging from viral particles to bacteria instigate excessive activation of CD4 T lymphocytes, macrophages and giant cells, which characterise GCA. Immune complex deposition causes an inflammatory reaction in the affected vessels. Inflammation starts in the adventitia and progresses to involve proliferation of the arterial media and intima. Endovascular damage ensues and the vascular lumen may become stenosed or completely occluded secondary to proliferation of the intima, and thrombosis. Ischaemic features and necrosis may develop in the areas supplied by the affected arteries.⁵⁻⁶ In our patient, the upper respiratory tract infection that preceded onset of the lingual numbness and pain may have triggered GCA. Her smoking likely contributed to poor wound healing and caused underlying arterial atherosclerosis, exacerbating ischaemic effects.

Diagnosis of GCA is initially clinical and later confirmed by histology on temporal artery biopsy. However, diagnosis can be hindered, clinically by the wide range of possible presenting symptoms. Histological changes can be patchy owing to the presence of skip lesions, such that diseased sections of vessel are inadvertently missed on biopsy, necessitating careful examination of the entire specimen at multiple levels.

American College of Rheumatology have stringent classification criteria for the diagnosis of GCA, a disease that necessitates long-term and possibly life-long steroid therapy, these being fraught with multiple side effects. The stipulated diagnostic criteria for GCA, also endorsed by the British Society of Rheumatology are age over 50 years, recent onset of headache, abnormal findings on

physical examination of the temporal artery (typically tender, nodular, pulseless and hard), raised ESR (>50 mm in the first hour), and a diagnostic temporal artery biopsy.⁹ A patient must meet three of these five criteria to be classified as having GCA. However, if the diagnosis of GCA is clinically suspected, the recommendation is for immediate start of the empirical high-dose corticosteroid therapy. Thus, empirical treatment should not be delayed by awaiting confirmation of diagnosis, by biopsy or with the aid of imaging, such as ultrasound or positron emission tomography scanning.⁹

Our patient did not volunteer any of the classical symptoms of GCA and only admitted to vague headaches and jaw pain on chewing upon repeated, specific questioning. Headache is the most commonly reported initial symptom, in approximately 70% of patients with GCA.⁸ Tongue claudication and tongue numbness is only present in 2-4% of patients and is very rarely the presenting symptom. However, there are a plethora of symptoms that patients with GCA complain of, including constitutional upset with fever and malaise, proximal myalgia (if there is concurrent polymyalgia rheumatica), visual disturbances including amaurosis fugax and diplopia, as well as scalp tenderness.⁸⁻⁹ Ischaemic sequelae of GCA are varied and may include lingual claudication and necrosis, necrosis of a periphery supplied by an end-artery, tissue gangrene of the scalp, bowel necrosis and myocardial infarction. Without appropriate clinical suspicion, these features may go unrecognised or be attributed to other conditions, especially in the absence of headache and temporal signs.⁹

Learning points

- Ischaemia of the tongue, a potentially reversible complication of giant cell arteritis (GCA), may be the only initial feature of underlying serious arteritis.
- GCA needs to be considered in elderly patients presenting with unexplained ischaemic symptoms.
- Prompt institution of appropriate therapy can lead to complete disease remission and preclude serious and irreversible end-organ damage, including blindness.
- We emphasise the importance of the admitting clinician maintaining a high index of suspicion of GCA and ensuring early multidisciplinary involvement, to avert preventable morbidity and mortality.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Kumarasinghe AP, Hepburn A, Reuther WJ, Pratt C. Temporal arteritis presenting with tongue necrosis. *BMJ Case Reports* 2012;10.1136/bcr-2012-007241, Published XXX

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