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ISCHEMIA OF THE TONGUE AND SCALP AS AN UNCOMMON PRESENTATION OF GIANT CELL ARTERITIS

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ABSTRACT

Giant cell arteritis (GCA) is a chronic large-vessel vasculitis that may present with systemic, neurological or ophthalmological manifestations. Lingual and scalp ischaemia are rare but can have severe potential complications that pose a diagnostic challenge and require immediate treatment to prevent irreversible outcomes.

We report the case of an 87-year-old woman who initially presented with holocranial headaches and tongue pain with partially detachable whitish plaques. She was diagnosed with GCA based on clinical features, elevated inflammatory markers and ultrasonographic findings of thickened temporal arteries. High-dose corticosteroid therapy was promptly initiated, leading to marked improvement of the reported symptoms. Nevertheless, on day 14 of hospitalisation, she developed septic shock from an abdominal source and subsequently died.

This case reports an atypical presentation of GCA – lingual and scalp ischaemia – emphasising the need to maintain a high index of suspicion even before these non-classical manifestations. A holistic clinical approach is essential to identify the various possible manifestations of this complex disease. Although tongue and/or scalp necrosis are rare complications, treatment remains aligned with current GCA guidelines. Most patients respond to corticosteroid therapy, or tocilizumab in cases of steroid-refractory lingual necrosis; non-responders are typically associated with a poorer prognosis.

KEYWORDS

Giant cell arteritis, vasculitis, tongue ischaemia, lingual necrosis, scalp necrosis

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LEARNING POINTS

- This report highlights the critical importance of considering giant cell arteritis (GCA) in the differential diagnosis of lingual and scalp ischaemia, enabling the exclusion of more common causes and ensuring the early initiation of appropriate treatment.
- The case emphasises the value of a multidisciplinary approach and continuous clinical surveillance, reinforcing the central role of the internal medicine physician in recognising atypical manifestations and coordinating follow-up in complex cases of systemic vasculitis.
- The report provides practical guidance for the diagnosis and management of rare presentations of GCA, offering evidence-based tools to support clinical decision-making and prevent irreversible complications.

INTRODUCTION

Giant cell arteritis (GCA), a granulomatous vasculitis with medium- and large-vessel involvement, is thought to be the most common form of vasculitis affecting individuals over 50 years of age. Immunologically, it is characterised by a mixed transmural inflammatory infiltrate causing the typical vascular lesions known as concentric rings. Dendritic cells activate CD4+ T cells, which differentiate into Th1 cells, stimulating macrophages and granuloma formation, leading to intimal hyperplasia and ultimately ischaemia of target organs. Macrophages release IL-6 and IL-1β, inducing Th17 cells, which sustain the inflammatory process. Th1-driven inflammation is associated with a lower response to glucocorticoids and a higher risk of relapse, whereas Th17-mediated responses demonstrate better treatment outcomes[1].

The diagnosis is established through clinical and laboratory criteria. According to the American College of Rheumatology, the presence of four out of five criteria is required: age over 50 years, reduced temporal artery pulse, new-onset headache, erythrocyte sedimentation rate greater than 50 mm/hour and an abnormal temporal artery biopsy[2].

Recently, the clinical paradigm of this disease has been redefined, being considered a multifaceted vasculitis disease that can involve cranial and extracranial arteries as well as the shoulder and hip girdles, either in isolation or combination. This heterogeneity often leads to delays in diagnosis configuring a diagnostic challenge and increases the risk of acute and chronic disease-related complications[1].

Persistent headaches, classically referred to the temporal region but also in other regions, are the most common presenting symptom, occurring in more than two-thirds of patients[3]. Cranial artery involvement is also associated with scalp tenderness, dysphagia, and mandibular and lingual claudication. In rare cases, occlusion of the temporal arteries or their branches may lead to necrosis of the scalp and tongue, ischaemic stroke and audiovestibular symptoms. Tongue alterations such as oedema, pallor, pain or intermittent claudication result from significant vascular involvement of the lingual artery – the first branch of the external carotid artery – since the extensive collateral network and multiple anastomoses usually protect this territory from ischaemia. Due to the tongue's rich vascular supply, bilateral lingual necrosis is even more uncommon. Non-specific systemic manifestations may also occur such as low-grade fever, fatigue and weight loss, making GCA one of the most common causes of fever of unknown origin in the elderly[1,3]. In addition to advanced age, other factors such as genetic variants in the major histocompatibility complex region, particularly HLA-DRB1*04:04, have been associated with disease development[1].

The diagnostic algorithm has also undergone significant changes. Although temporal artery biopsy has long been the only diagnostic tool, non-invasive imaging modalities such as vascular ultrasound, angiography, FDG-PET and magnetic resonance imaging have gained prominence and now complement the diagnosis of GCA, but they may also fail in some cases[1].

Tongue necrosis and vision loss associated with GCA are irreversible complications that can lead to devastating outcomes – permanent vision loss or tongue amputation. Therefore, high-dose glucocorticoids should be initiated as soon as GCA is suspected, even before biopsy yields the definitive diagnosis. Glucocorticoids should be maintained until symptoms subside or inflammatory markers normalise. Relapses are common in GCA as corticosteroids are gradually tapered; thus, prolonged tapering is often necessary. Unfortunately, patients are at risk of glucocorticoid toxicity or may exhibit glucocorticoid resistance, as well as increased risks of infection, cardiovascular disease, osteoporosis, osteonecrosis or diabetes. Although research does not support the efficacy of many immunosuppressants, interleukin-6 (IL-6) inhibitors have become a highly relevant therapeutic option following evidence of the efficacy of tocilizumab, resulting in better disease control, fewer relapses and facilitating corticosteroid tapering. New promising drugs are under investigation and are expected to strengthen the therapeutic arsenal available for GCA[1,3,4].

CASE DESCRIPTION

An 87-year-old woman with a medical history of anticoagulated atrial flutter, diabetes mellitus and hypertension presented to the emergency department with holocranial headaches refractory to analgesics. She also reported tongue pain with partially detachable whitish plaques, accompanied by a four-week history of

progressively worsening dysphagia for solid foods and dysphonia (hoarse voice).

She had been previously evaluated at the emergency department for the same symptoms one week earlier and was treated with oral fluconazole and topical nystatin without improvement. She was apyretic and denied visual symptoms, had no allergies or history of oral trauma. Vital signs on admission were within normal limits.

On physical examination, the tongue exhibited an extensive ulcerated lesion occupying the transition between the anterior and middle thirds, covered by a painful, yellowish, necrotic-appearing plaque, with irregular and indurated borders and reduced mobility *(Fig. 1)*. Additionally, painful necrotic lesions were observed on the scalp *(Fig. 2)*, along with multiple tender and mobile submandibular lymphadenopathies. Bilateral temporal artery pulses were absent.



Figure 1. Tongue with an extensive ulcerated lesion occupying the transition between the anterior and middle thirds, covered by a yellowish necrotic-appearing plaque. The lesion is painful with irregular, indurated borders.



Figure 2. Scalp lesions consistent with scalp ischaemia.

Laboratory tests showed a white blood cell count at 11.6 $\times 10^9$/l (reference range 4–11 $\times 10^9$/l), haemoglobin at 9.8 g/dl (13–18 g/dl), and platelets count at 580 $\times 10^9$/l (150–400 $\times 10^9$/l). Inflammatory markers were elevated, with C-reactive protein at 109 mg/l (<3 mg/l) and erythrocyte sedimentation rate at 116 mm/h (0–29 mm/h). Autoimmune screening (ANCA, ANA, ENA, anti-SSA, anti-SSB and anti-dsDNA) was negative.

A Doppler ultrasound of the neck and transcranial vessels revealed signs of proximal occlusion of the superficial temporal arteries. Given the strong suspicion of GCA, high-dose corticosteroid therapy was initiated (methylprednisolone 500 mg daily for three days, followed by prednisolone 1 mg/kg–60 mg). A nasogastric tube was placed for feeding and medication administration due to severe dysphagia. Further investigation included an incisional biopsy of the ulcerated lesion, which showed ulceration with granulation tissue. A temporal artery biopsy revealed lymphohistiocytic infiltrate of the media layer, intimal thickening and giant cells consistent with GCA. No signs of malignancy were observed. Fungal elements were not identified on staining, and immunohistochemical markers were negative for cytomegalovirus, herpes simplex virus and varicella-zoster virus.

A cervical computed tomography angiography demonstrated a discontinuity on the dorsal aspect of the tongue at the midline, lateralised to the right, measuring approximately 13 × 25 × 11 mm, corresponding to the ulcerated lesion observed on physical examination. No focal arterial occlusion was noted; however, decreased vascularisation was observed in the lingual arteries.

A CT angiography of the cerebral arteries showed no signs suggestive of aortitis or vasculitis. A thoraco-abdomino-pelvic CT scan and transthoracic echocardiogram were negative for large-vessel involvement. Fundoscopic examination by an ophthalmologist revealed no optic nerve abnormalities, oedema or haemorrhage.

Over the following week after starting treatment, gradual improvement was noted in mandibular claudication and headache, along with a reduction of necrotic tissue beneath the tongue and decreasing inflammatory markers. However, on day 14, the patient developed septic shock from an abdominal source, leading to multi-organ failure and subsequent death.

DISCUSSION

Tongue ischaemia constitutes an atypical and rarely described presentation in the literature on GCA, often responsible for

delays in diagnosis and treatment. Patients over 50 years old presenting with tongue pain, pallor or oedema, unresponsive to treatment for infection or angioedema, should raise suspicion of an ongoing vasculitic process. Tongue ischaemia as a manifestation of GCA was first reported in 1959 and has been documented since then[4]. It occurs in approximately 1% of cases as an initial presentation[5], but appears to be a late finding affecting around 25% of patients, most of whom are elderly women[4]. Progression to tongue necrosis is a marker of poor prognosis, being associated with increased mortality, although it is not included among classification criteria[6]. Due to the rich vascular supply from branches of the lingual, facial, pharyngeal and palatine arteries, unilateral presentation of lingual necrosis is more common than bilateral involvement[4-6].

Clinical manifestations of tongue involvement in GCA entails a spectrum of signs and symptoms ranging from claudication, blanching, oedema, glossitis, pain, cyanosis and erythema, eventually progressing to necrosis. Delayed diagnosis and treatment may lead to severe complications such as autoamputation of necrotic tissue, aspiration and respiratory failure[5]. When evaluating tongue necrosis, several important differential diagnoses must be considered. These include underlying malignancy, adverse drug reactions, exposure to radiotherapy and chemotherapy, cardiovascular aetiologies (haemorrhage, embolism and post-cardiac arrest), infections such as syphilis, tuberculosis or herpes, and systemic vasculitis (GCA and ANCA-associated vasculitis), with the latter being the most likely diagnostic hypothesis[4].

Scalp necrosis in the context of GCA is generally a sign of active disease involving multiple blood vessels and has been associated with a higher incidence of visual complications. It is also associated with an increased incidence of tongue necrosis, and both manifestations may occur simultaneously. Concurrent necrosis of the tongue and scalp has been previously reported in only two cases in the literature[7].

When evaluating a suspected diagnosis, biopsy may have a variable diagnostic yield. In addition to clinical assessment and laboratory investigations, the diagnosis of GCA can be further supported by colour Doppler ultrasonography, as was demonstrated in our case[4,8].

A lower threshold for considering this entity and a prompt treatment with high-dose corticosteroids are crucial to prevent devastating outcomes such as tongue amputation or permanent visual loss. Most patients respond well to corticosteroids (with doses ranging from prednisolone 1 mg/kg to methylprednisolone 1,000 mg); conversely, steroid-sparing agents such as methotrexate, azathioprine, cyclophosphamide and anti-TNF therapies have shown limited efficacy[7,8].

The initial intensity of the inflammatory response in GCA can be determined by five parameters: platelets > 400,000/mm^3, temperature > 37.5°C, leukocytes > 11,000/mm^3, ESR > 100 mm/h and haemoglobin < 11 g/dl. Patients who meet at least four of these criteria are considered at high risk for recurrence while tapering treatment and corticosteroid dependence[8].

In corticosteroid-refractory cases, tocilizumab has shown promising results and is considered by some authors in the literature as the preferred disease-modifying antirheumatic drug. Three cases requiring tongue amputation have been reported. One case described a favourable response to combined corticosteroid and tocilizumab therapy as first-line treatment, due to extensive arterial bed involvement supported by abnormal ultrasonographic findings of the deep lingual artery, namely halo sign, increased intima-media thickness and markedly reduced blood flow[1,4]. Eight cases in the English literature presented with initial tongue necrosis, leading to delayed diagnosis and subsequent blindness[4,5]. Depending on the extent of necrosis, patients may require amputation or surgical debridement of the necrotic area[1,4].

This case has some limitations that can be pointed out, such as the inability to initiate treatment with other immunosuppressants, as symptoms worsened rapidly over a two-week period. The patient's death may be attributed to infectious complications related to high-dose corticosteroid therapy – particularly considering her advanced age – or to disease progression with occlusion of aortic branches. It remains unclear whether early initiation of tocilizumab would have been a better option or even improved the overall prognosis.

In summary, GCA is characterised by a heterogeneous presentation, which may include atypical manifestations such as tongue ischaemia. This report highlights the importance of early diagnosis and treatment, as well as the need to maintain a high index of suspicion for potential complications that may develop along the course of the disease, even under corticosteroid therapy. Prompt diagnosis and appropriate treatment of GCA are crucial to prevent progression of tongue necrosis; notwithstanding this, the adverse effects of high dose corticotherapy must considered. The authors recommend that any patient over 50 years of age presenting with lingual ischaemia of unknown cause should undergo urgent investigation for GCA.

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