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DOCTORS IN TRAINING

The Importance of Early Recognition of Rare Ischaemic Complications of GCA to Prevent Permanent Vision Loss

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Abstract

Scalp and tongue necrosis are rare ischemic manifestations of giant cell arteritis (GCA). Early recognition of these conditions is crucial to preventing permanent visual loss (PVL). We present two cases of scalp and tongue necrosis, respectively, where a delay in diagnosis resulted in irreversible vision loss and severe complications. These cases highlight the importance of educating non-rheumatologists about these manifestations to ensure prompt steroid treatment, which can prevent vision loss and reduce morbidity in GCA patients.

Key words: scalp necrosis; tongue necrosis; giant cell arteritis

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Introduction

Ischemic manifestations of giant cell arteritis (GCA) due to reduced blood flow leading to tongue and scalp necrosis, are rare presentations of this vasculitis (Prieto-Peña et al, 2021). A higher incidence of permanent visual loss (PVL), the most feared complication of GCA, is associated with these presentations compared to other common symptoms of GCA (Brodmann et al, 2009; Tsianakas et al, 2009). The delay in diagnosis contributes to significant morbidity and mortality in this group of patients.

We present two cases to underscore the importance of early recognition of scalp and tongue necrosis to prevent vision loss. There is a need to educate non-rheumatologists about these manifestations to avoid any delay in management.

Case 1

An 87-year-old lady presented with 3-day history of worsening confusion and hallucinations. She also reported double vision for the last 2 weeks and visual loss in both eyes for the last 1 week. Her medical history included generalized osteoarthritis, hypertension, hypothyroidism, asthma, and previous herpes zoster infection. Additionally, she had a history of headaches and a weight loss of 2–3 stones over the last 6–7 weeks.

Upon reviewing her clinical record, it was noted that she had visited her general physician and also attended the Emergency Department (ED) twice within 6 weeks

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before admission, presenting with a headache and a painful lump over the scalp. Her C-reactive protein (CRP) levels during the ED attendances were 336 mg/L (normal range: 0–5 mg/L) and 31 mg/L respectively. There was no clear cause of scalp lesions mentioned in the clinical record.

The rheumatology team, during the current admission, identified scalp lesions consistent with scalp necrosis (Fig. 1). The visual hallucinations were likely Charles Bonnet phenomenon secondary to visual loss. Her CRP was 33 mg/L on admission. Further questioning revealed that she had jaw claudication and diplopia before the vision loss. She was initiated on oral prednisolone 60 mg daily. An ophthalmology review indicated bilateral optic disc swelling and poor visual acuity. She could only perceive hand movements in the right eye and light perception in the left eye. The temporal artery biopsy confirmed the diagnosis of active GCA. Unfortunately, there is no improvement in her vision after starting steroids and she was registered blind.

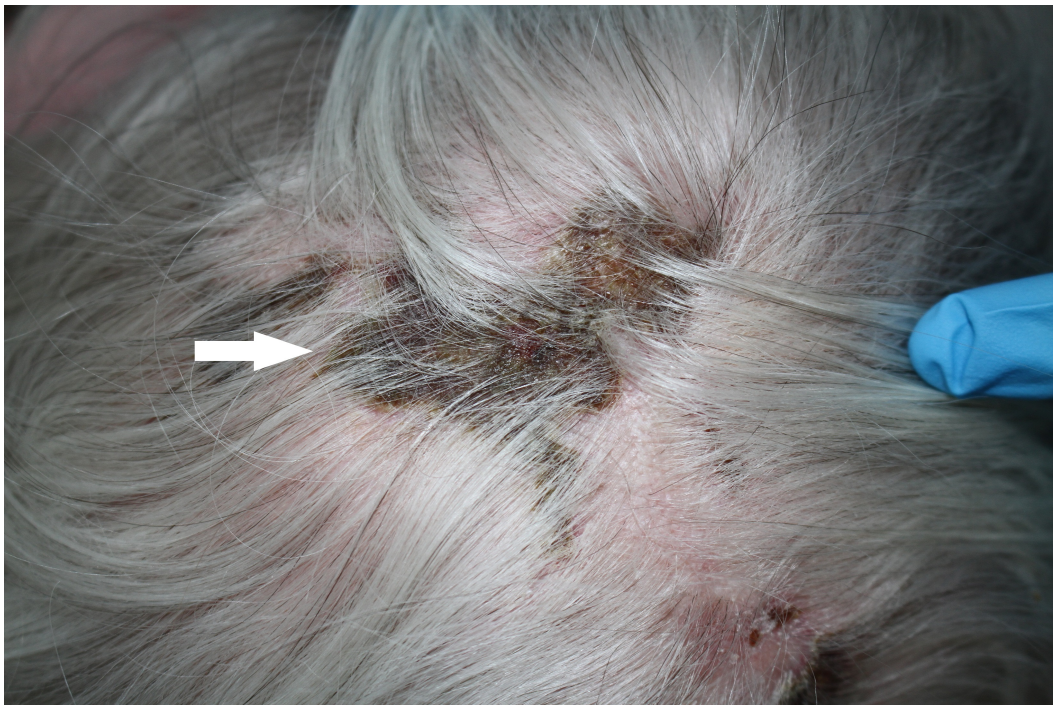

Fig. 1. Scalp necrosis (indicated by the white arrow).

Case 2

A 75-year-old lady admitted under the Ear, Nose, and Throat (ENT) team presented with 1-week history of a sore tongue causing dysphagia, dysphonia, and claudication on movement. She had a medical history of gastro-oesophageal reflux disease. Further questioning revealed blurred vision in the left eye, headache and a weight loss of up to 3 stones in the last 6 months.

Clinical examination showed a tender, dusky/bluish discoloured tongue with overlying slough (Fig. 2). The complete blood count was fairly unremarkable.

Nasendoscopy and positron emission tomography/computed tomography (PET/CT) of the head and neck did not reveal an obvious cause for the symptoms. She was mainly treated with nystatin and metronidazole for a suspected infection during her 3-week hospital stay.

A month later, she presented to the eye casualty with a sudden loss of vision in both eyes, along with pain in the right temple and jaw claudication. Fundoscopy examination indicated ischemia of the right optic disc and blood tests showed a CRP level of 60 mg/L and an erythrocyte sedimentation rate (ESR) of 27 mm/h (normal range: 0–30 mm/h). Intravenous (IV) pulsed methylprednisolone was initiated with a possible diagnosis of GCA, which was later switched to oral prednisolone. Her temporal artery biopsy confirmed the diagnosis of active GCA. Similar to Case 1, her vision did not improve after starting steroids. She readmitted with aspiration pneumonia and developed left upper limb ischemia secondary to occlusion of the mid-left brachial artery. She sadly passed away due to these complications.

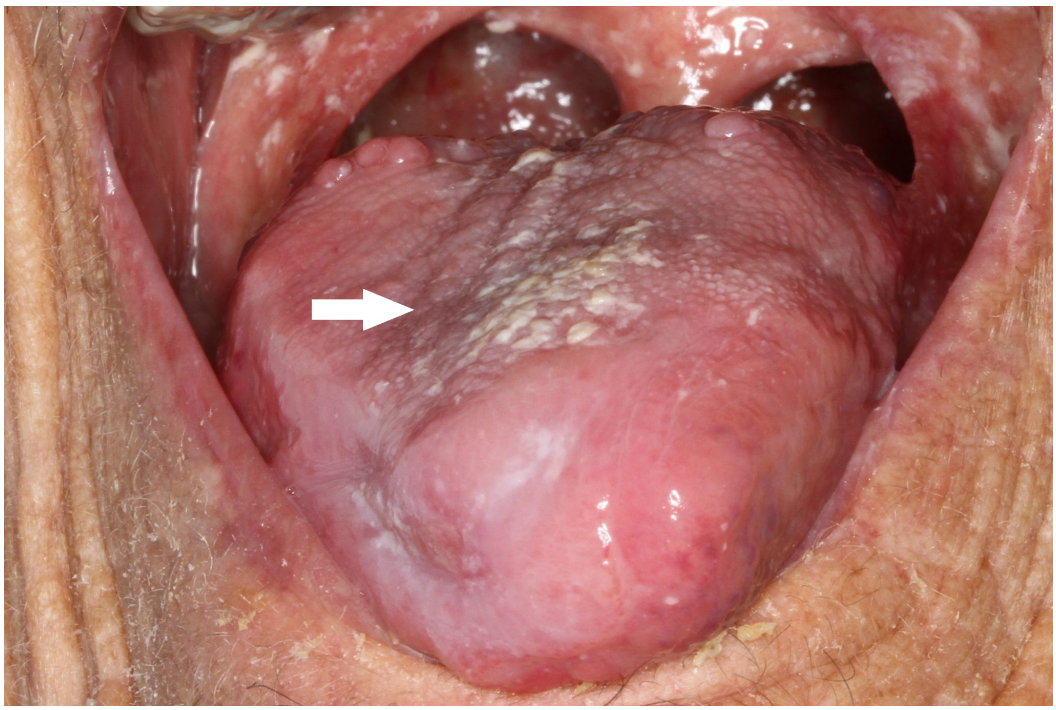

Fig. 2. Tongue after sloughing of necrotic tissue on the right side (indicated by the white arrow).

Discussion

It is noticed in both cases that there was a significant delay of more than a month before the final diagnosis was made. Visual symptoms for GCA can be transient or permanent, ranging from blurring of vision, diplopia and amaurosis fugax. Once permanent visual loss is established, it is less likely to be reversible. Up to 17% of GCA patients develop permanent visual loss (Mackie et al, 2011). There is higher incidence of PVL associated with scalp or tongue necrosis in GCA

patients. Tsianakas et al (2009) describe that GCA patients with scalp necrosis have a 32% incidence of visual loss compared to 20% of patients without scalp necrosis. Kitamura et al (2020) did systemic review where GCA found to be the single most common cause of tongue necrosis.

There is no specific diagnostic criterion available for diagnosing GCA. However, the 2022 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for GCA highlight important points in the history that need to be considered in suspected GCA patients (Ponte et al, 2022). Although a temporal artery biopsy is the gold standard investigation to confirm the diagnosis of GCA, however, doppler ultrasound for temporal +/– axillary artery has become an integral part of rapid access GCA pathways in large tertiary centres. Both of these investigations have their limitations in terms of the risk of low yield after starting steroids. Therefore, timely referral to experts in diagnosing GCA is very important to ensure a correct diagnosis (Mackie et al, 2020).

Suspected GCA patients warrant urgent referral to rheumatology services, but treatment with systemic steroids should be started without any delay. The initial glucocorticoid dose is oral prednisolone 40–60 mg daily. However, if there are ischemic visual manifestations in suspected GCA patients, they should be referred to an acute hospital for urgent (same-day) ophthalmology assessment and IV methylprednisolone. The dose for IV methylprednisolone is 500–1000 mg daily for 3 days, followed by a switch to oral prednisolone. If there is a delay in obtaining IV methylprednisolone, treatment with oral prednisolone at a dose of 60–100 mg daily should be started immediately (Mackie et al, 2020).

Conclusion

These cases emphasize the significance of early diagnosis of GCA when patients present with scalp and tongue necrosis. Both conditions are linked to a higher incidence of mortality in GCA patients (Chehem Daoud Chehem et al, 2024). The majority of these patients ended up seeing non-rheumatologists as their ischemic symptoms raised the possibility of underlying malignancy. Timely diagnosis and early treatment with steroids would help save vision and prevent further morbidity in this vulnerable group of patients.

Key Points

- Scalp and tongue necrosis can be the presenting symptoms of GCA and delay in diagnosis can lead to irreversible vision and loss.
- Early treatment with steroids and referral to Rheumatology for confirmation of the diagnosis is crucial to prevent vision loss.
- These patients often get referred to other medical specialities with a suspicion of malignancy and there should be more awareness of these rare presentations of GCA among the wider medical community.

Curriculum Checklist

This article addresses the following requirements from the general internal medicine training curriculum:

- Managing rheumatological emergencies
- Large vessel vasculitis
- Teaching and training
- Research

Availability of Data and Materials

All data and materials of this study are included in this article.

Author Contributions

MTG is the first author. DR and DM are co-authors. MTG and DM contributed for conception and design of manuscript. MTG, DR and DM did literature review and data collection & interpretation. MTG and DR involved in drafting manuscript. MTG and DM revised the manuscript to add important points. All authors contributed to important editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

The written consent was taken from Case 1. Unfortunately, Case 2 died and there was no family member to take consent on her behalf.

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Conflict of Interest

The authors declare no conflict of interest.

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