CASE REPORT

Giant cell arteritis presenting as tongue necrosis

Ritesh Kohli, Eleni Tiniakou, Joao M Nascimento, Gbonjubola Oyefeso

ABSTRACT

Introduction: Giant cell arteritis (GCA) is a large and medium-vessel systemic vasculitis affecting predominantly the internal and external carotid arteries with particular affinity for its extracranial branches (especially, the superficial temporal artery). The most frequent complication of this disorder is visual loss. We report the case of a patient who suffered tongue necrosis (a rare complication) secondary to giant cell arteritis. Case Report: A 71-year-old female who after having her dentures refitted developed severe pain, edema of the tongue, odynophagia, dysphagia and resultant anorexia. She was admitted with the diagnosis of glossitis and initially treated with antifungals and later antiviral medications without improvement. She subsequently developed tongue ulcerations, severe pain in temporomandibular joint (TMJ) with right sided headache. Temporal arteritis was suspected at this point, re-examination revealed palpable and tender temporal arteries (R>L). Tongue examination demonstrated dark greyish plaques in the dorsum with areas of ulceration without palatal involvement. An elevated ESR and the right sided temporal artery biopsy that followed demonstrating chronic inflammation with the presence of giant cells confirmed the diagnosis of GCA. Treatment with oral prednisone (1 mg/kg/day) produced a rapid improvement of her symptoms and ulcers with complete resolution at three months. Conclusion: The presence of glossitis in GCA represents a severity marker of this disease and is associated with a higher risk of vision loss and mortality. As such a high level of suspicion is necessary for the early recognition of this rare presentation and prompt institution of treatment to prevent the aforementioned complications.

Keywords: Giant cell arteritis (GCA), Scalp necrosis, Temporal artery biopsy

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INTRODUCTION

Giant cell arteritis (GCA) is a systemic vasculitis affecting large and medium sized arteries. It chiefly involves the extra cranial branches of carotid artery, particularly the posterior ciliary arteries that supply the optic nerve and the superficial temporal artery. The most frequent complication of this disorder is visual loss. We report a case of a patient who suffered rare complication, tongue necrosis, as a result of GCA. Early diagnosis and treatment of this can limit tissue injury and the progression of disease.
CASE REPORT

A 71-year-old female who after having her dentures refitted developed severe pain and edema of the tongue. She was admitted with the diagnosis of glossitis and initially started on antifungal therapy. She later developed tongue ulcerations and was started on antiviral medications with no significant improvement. Subsequently, she developed severe pain in temporomandibular joint (TMJ) and right sided headache to the extent that she had marked difficulty swallowing and decreased oral intake. Her past medical history was positive for well controlled hypertension and hyperlipidemia alone. Her social and occupational history was non-contributory as were the review of systems. Temporal arteritis was suspected at that point and re-examination revealed palpable and tender temporal arteries especially on the right side. The oropharyngeal examination revealed the presence of dark-greyish plaques and focal areas of ulceration on the dorsum of the tongue. Laboratory examination was remarkable for a grossly elevated sedimentation rate at 95. A right sided temporal artery biopsy was performed and demonstrated chronic inflammation and the presence of giant cells confirmatory for the diagnosis of GCA.

Treatment with oral prednisone (1 mg/kg/day) produced a rapid improvement of her symptoms and a gradual improvement in the appearance of the ulcerated areas. The lesions were completely resolved at a three-month follow-up.

DISCUSSION

Giant cell arteritis is a chronic granulomatous vasculitis, of unknown etiology, that affects individuals older than 50, especially females. Aging is the single greatest risk factor for the disease. The GCA never occurs before the age of 50 years. Although the cause of GCA is unknown, the disease appears to be T cell dependent and antigen driven [1].

The classic manifestations of GCA are headache, jaw claudication, polymyalgia rheumatica and visual symptoms including diplopia and loss of vision. Atypical manifestations include dry cough, pinna or parotid gland pain due to involvement of posterior auricular artery, fever of unknown origin and otolaryngeal manifestations including tongue pain, glossitis, dental pain, tongue ulceration and gangrene. Necrosis of the tongue occurs rarely owing to the extensive collateral blood supply. There is usually associated dysphagia, excessive salivation, tongue pain and masticatory claudication prior to a diagnosis of arteritis [2]. These symptoms are important warning signs of impending lingual infarction.

The diagnosis of GCA is mostly clinical. Recently, the American College of Rheumatology, established diagnostic criteria, which states that to be classified as having a GCA, a patient must meet three of the following five criteria.

(i) Aged 50 years or older.

(ii) The presence of new onset localized headache.

(iii) Temporal abnormalities like temporal tenderness or decreased temporal pulse.

(iv) ESR of 50 mm or higher.

(v) Abnormal temporal artery biopsy findings demonstrating mononuclear infiltration or granulomatous inflammation [3].

The presence of three or more of these five criteria is associated with a sensitivity of 93.55% and a specificity of 91.2%.

In clinical practice, establishing the diagnosis of GCA requires a biopsy of the temporal artery. The temporal artery biopsy must be carried out in all patients with a suspicion of GCA, even if the ESR has been normal. If the biopsy is negative, and the clinical suspicion is high, contralateral biopsy should be performed. Temporal artery biopsy is positive in only half of the patients who manifest the disease, as skip lesions occur commonly so a negative biopsy does not exclude GCA. The administration of corticosteroid given for less than two weeks does not reduce the yield of temporal artery biopsy [4]. In cases of lingual necrosis, tongue biopsy is usually nonspecific and is not indicated.

The most dreaded consequence of GCA is visual loss, which is usually irreversible. The early treatment of patients with GCA is crucial to avoid visual loss and the therapy should be started based on clinical suspicion, and not delayed for the biopsy results. Once the diagnosis of GCA is suspected, early treatment with steroids is recommended, to prevent complications and promote the complete resolution of this self-limiting condition. Typically, the treatment begins with prednisone 1 mg/kg body weight in adults [5]. It must be maintained until the symptoms have disappeared and the ESR is back to normal.

In the absence of characteristic signs and symptoms, diagnosing GCA can pose a challenge. This case reinforces the importance of suspecting GCA as cause of tongue necrosis especially in the absence of typical symptoms like headache and vision loss. Tongue necrosis is a rare occurrence in GCA and its presence represents a severity marker of the disease associated with an elevated risk of vision loss as well as overall higher mortality rates, in comparison to patients not presenting with this manifestation.

CONCLUSION

The aim of this article is to alert the physicians to the atypical manifestations seen in this disease like tongue claudication and tongue infarction which can all serve as a warning sign, as such a high level of suspicion must be held for these clinical findings, in order to initiate prompt and proper treatment and avoid blindness.

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Author Contributions
Ritesh Kohli – Substantial contributions to conception and design, Acquisition of data, Analysis and
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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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