Tongue Ulceration and Necrosis in Giant Cell Arteritis: A Case Report

Nathan M. Schularick, MD; Raymond W. Kung, BS; J. Robert Schieffarth, MD; Henry T. Hoffman, MD
Department of Otolaryngology, University of Iowa Hospitals and Clinics

Abstract

Objectives: This study aims to facilitate diagnosis of Giant Cell Arteritis (GCA) by reporting its less known symptoms of ulcerative tongue necrosis and non-throbbing extra-temporal headache. This study also serves to alert the surgeon to the risk of frontalis palsy with temporal artery biopsy in the danger zone.

Methods: A 66-year-old Caucasian female presented to the ER with a painful, burning, non-healing tongue ulcer with concurrent episodes of transient right-sided vision loss. She reported a one-month history of non-pulsatile, predominantly occipital headache with scalp tenderness along the “veins” while combing her hair. She was found to have elevated LFTs and ESR.

Results: The patient was started on Solumedrol followed by Prednisone taper therapy for presumptive GCA, later confirmed by right temporal artery biopsy. The tongue ulceration identified at the time of diagnosis had healed 10 days after treatment was instituted. Her headaches and vision loss also resolved without sequelae. No frontalis weakness was noted at her most recent follow up.

Conclusions: Tongue ulceration with necrosis, a rare complication of GCA, can be its first symptom. GCA does not always consist of throbbing temporal headaches and should be included in the differential for elderly patients presenting with headaches of any characteristics. Surgeons performing temporal artery biopsies should take into consideration the potential course of the frontal branch to prevent frontalis palsy.

Introduction

Giant cell arteritis (GCA), also known as temporal arteritis, is a chronic systemic vasculitis of medium and large sized arteries. GCA is one of the most common forms of vasculitis in adults. It almost exclusively affects white individuals 50 years and older. Though it occurs in only about 30% of cases. Constitutional symptoms of fatigue, malaise, fever, anorexia, weight loss, and night sweats are also common. GCA is also highly associated with polymyalgia rheumatica, and when present, may be associated with neck, shoulder or hip pain and/or stiffness.

Physical exam may reveal tenderness of the scalp or temporal arteries and decreased temporal artery pulses. Serum studies often detect elevated ESR, CRP, and LFTs (especially Alkaline Phosphatase) as well. Duplex sonography may reveal hypoechoic circumferential arterial wall thickening, termed the halo sign. Rarely, GCA may be complicated by tongue ulceration and necrosis as is presented in this case.

Case Report

A 66-year-old Caucasian female presented to the Emergency Department (ED) for multiple episodes of transient right-sided vision loss. She had a five day history of a painful, burning, non-healing tongue ulceration (Figure 1). She reported a one-month history of intermittent, non-pulsatile, predominantly occipital headache with scalp tenderness along the “veins” while combing her hair.

Two weeks before presentation to the ED, she had a total thyroidectomy with neck dissection for papillary thyroid carcinoma metastatic to the cervical lymph nodes. In the ED, she was found to have elevated ESR, CRP, and LFTs. The patient was immediately started on 1000mg of IV pulse methylprednisolone for a presumptive diagnosis of GCA. A temporal artery biopsy was performed two days later.

Histological analysis of a right superficial temporal artery biopsy revealed inflammatory infiltrate of the tunica media and adventitia, disruption of the elastic lamina, and granulomatous change, confirming the diagnosis (Figures 2 and 3). She was maintained on 1000mg of methylprednisolone daily for three days followed by a prednisone taper of 60mg daily for one week, 70mg daily for two weeks, then 60mg daily for two weeks. Ten days after initial presentation the ulcer had healed as a 0.5x1.0cm defect at the tongue tip (Figure 1). No frontalis weakness was noted at her most recent follow up.

Discussion

Patients with GCA may initially present to otorhinolaryngologists. Prompt recognition of the symptoms of GCA is crucial to decrease the risk of rapid progression to blindness or stroke. Establishing the diagnosis of GCA can be challenging due to the wide range of clinical symptoms and the large number of different blood vessels. Lingual ulceration and necrosis can be the initial presentation. In addition to systemic steroids, reports describe managing lingual necrosis with debridement and healing by secondary intention. Rocky et al. recommend resection and primary closure to reduce potential scarring when larger areas of necrosis are encountered.

Although the headache of GCA is classically described as severe, throbbing, temporally located, and acute in onset, the pain is often nonspecific in characteristic. The patient in this case reported a chronic non-throbbing occipital headache of mild severity. GCA should therefore be considered in the differential for elderly patients presenting with new onset headache – even when not as classically described.

The gold standard for the diagnosis GCA is histologic confirmation by arterial biopsy. Temporal arteritis is most commonly chosen due to its accessibility and frequency of involvement. Initial planning may include doppler mapping the course of the artery prior to incision, with special consideration of the potential course of the frontal branch of CN VII to prevent frontalis palsy. Biopsies taken from within Bhatia’s “danger zone” (Figure 4) may result in brown piosis due to the proximity of the frontal branch of the superficial temporal artery to the frontal branch of CN VII. Occasionally, the frontal branch of CN VII may cross the superficial temporal artery at an unusually high location (Figure 5) To reduce the risk of forehead paralysis, several strategies are suggested. Yoon suggests the surgeon biopsy the parietal rather than the frontal branch of the superficial temporal artery Bhatia et al. recommends dissection of the artery go no deeper than the superficial temporal fossa, as the facial nerve runs just deep to this within the loose areolar layer.

Systemic steroids remain the only means to prevent irreversible visual loss in GCA. Treatment should be started immediately upon strong clinical suspicion of giant cell arteritis. Smith et al reported that steroids will not obscure histologic examination if the biopsy is completed within two weeks of the start of medication. The European League Against Rheumatism (EULAR) currently recommends an initial dose of prednisone at 1 mg/kg, not to exceed 60 mg a day, for four weeks, followed by gradual tapering. If severe ischemic complications such as visual loss or stroke are present, 0.5 to 1g of an intravenous bolus of methylprednisolone is recommended initially and each of the next three days. Conversion to oral steroid with management by a tapering dose based on the clinical picture is recommended. In the presence of chronic disease, prednisone should reach 10 to 15 mg daily by month 3, and 5 mg daily by month 6 after diagnosis. Methylprednisolone/prednisone should then be maintained at 5 mg daily for at least 12 months.

Conclusion

Prompt diagnosis and treatment of giant cell arteritis is useful to diminish the risk of disease progression including vision loss and stroke. Tongue ulceration with necrosis is a rare complication of GCA but can be a presenting symptom. Because the headache of GCA may be nonspecific, arteritis should be included in the differential diagnosis of new onset headache occurring in the elderly. The risk of iatrogenic frontalis palsy in the course of performing a temporal artery biopsy may be diminished by either sampling the parietal rather than the frontal branch of the superficial temporal artery, or by limiting incisions to the superficial temporal fossa for arterial biopsy.

Contact
Nathan M. Schularick, M.D.  schularickn@healthcare.iowa.uiowa.edu
Raymond W. Kung, B.S.  raykung68@gmail.com

References