An Atypical Presentatio n of Giant Cell Arteritis

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An Atypical Presentation of Giant Cell Arteritis

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HISTORY OF PRESENTING ILLNESS:

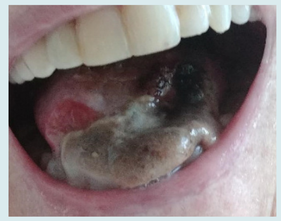
An 87-year-old female with history of polymyalgia rheumatica (PMR), osteoporosis, mild cognitive impairment, insomnia, dyslipidemia, and GERD presented to the hospital in June 2018 from her nursing home with complaints of new onset left jaw pain, dysphagia, and tongue pain for 1 week. Two and a half weeks prior to hospitalization, she was evaluated by her primary care provider due to symptoms of painful, sore shoulders with associated new ambulatory difficulties in the setting of an elevated ESR of 68. She was subsequently treated with prednisone 25 mg/d titrated down to 10 mg/d. Her symptoms persisted and she presented to the hospital. On admission, labs were unremarkable except a WBC of 14.3. CT imaging showed no acute inflammatory process in maxillofacial region and CXR was suspicious for left lingular pneumonia which was treated with broad spectrum antibiotics. Her tongue lesion was initially evaluated by dentistry who felt the etiology was likely due to oral or esophageal candidiasis that resulted after prednisone use. She was treated with magic mouthwash and nystatin without resolution. She was subsequently seen by Ear, Nose, and Throat and Infectious Diseases consultants who felt the tongue ulceration and ecchymosis was due to Herpes Simplex I (HSV-1). She was empirically treated with acyclovir and viscous lidocaine. Her hospitalization was complicated by failure of swallow study secondary to tongue pain and Dobhoff placement was required. She was discharged on her outpatient prednisone dose of 10 mg/d.

After hospitalization, the patient was referred to Rheumatology and seen on 7/18/18 for concern of an atypical presentation of giant cell arteritis. Her tongue pain was improved but not resolved and her PMR symptoms were controlled. She denied double vision, blurry vision, headache, temporal head tenderness, scalp tenderness and sensitivity. Her ESR/CRP were serially monitored to monitor progression of disease (see table 1). Due to persistently elevated ESR/CRP in the setting of PMR with unresolved tongue lesion, the patient was referred to Vascular Surgery for temporal artery biopsy. She underwent bilateral temporal artery biopsy on 9/4/18 with results consistent with temporal arteritis. The patient's dose of prednisone was increased to 40 mg/d and later to 50 mg/d with methotrexate for steroid sparing due to persistently elevated CRP. After treatment with adequate medication doses, the patient's tongue lesion improved and inflammatory markers decreased.

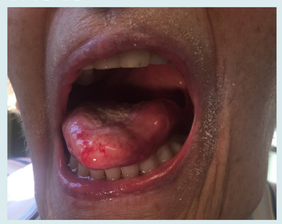
Table 1. Trend of CRP and ESR

	5/25/18	7/13/18	8/22/18	9/17/18	10/8/18	12/14/18	1/14/19
ESR (mm/hr)	68	71	72				
CRP (mg/L)		44.8	35.9	4.8	45.7	12.9	<3.0

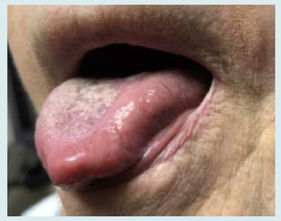
6/29/18



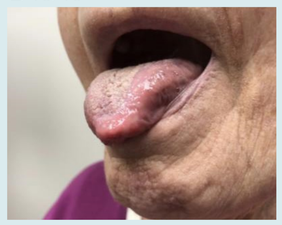
7/18/18



12/12/19



2/19/19



DISCUSSION:

In this case, we describe a patient with polymyalgia rheumatica (PMR) who presented with a necrotic tongue ulcer as her main manifestation of giant cell temporal arteritis (GCA). Demographics from a systemic review of twenty-five patients with GCA and tongue necrosis suggests average age at diagnosis of 77 and 23 of 25 were female.[1] In a British cohort of patients with GCA the presence of tongue and scalp necrosis were manifestations of more severe GCA and had higher incidence of visual loss.[2] Other atypical features have been reported including cough, trismus, hearing loss, and facial swelling.[3,4,5,6,7]

Patients with GCA present frequently with headaches (90%), polymyalgia rheumatica (34%), jaw claudication (50%), and blurred vision (40%).[8] Other findings include fever, elevated sedimentation rate, and leukocytosis. Ocular symptoms are notably the most devastating and treatment should be prompted immediately if suspicion for disease is high. Bilateral temporal artery biopsy has been associated with a >90% negative predictive value from retrospective data at Mayo Clinic[9] though this degree of diagnostic accuracy is suspected to be much lower[10]. GCA can have devastating consequences including stroke and irreversible blindness if not treated early. The use of corticosteroids continues to be the standard of care. The duration of treatment depends upon tolerability of steroid taper. Adjunctive therapies such as methotrexate, azathioprine, dapsone, and anti-TNF-alpha agents have been of limited proven benefit.[11] The use of tocilizumab, an anti-IL-6 agent, along with steroid taper has shown early promising outcomes.[12]

CONCLUSION:

Tongue necrosis is not part of the criteria cited by the American College of Rheumatology for GCA. [13] However, its presence in a patient with elevated ESR >50mm/hr with age >50 warrants referral to vascular surgery for temporal artery biopsy. Early detection of this atypical manifestation can lead to early diagnosis and treatment as well reduction of morbidity.

REFERENCES:

1) https://www.hindawi.com/journals/crim/2017/6327437/tab1/
2) Rheumatology (1997). 36:814-816.
3) Clin Exp Rheumatol. 2015; 33: S164–S170.
4) HNO. 1984; 32: 115–118.
5) Am J Otolaryngol. 2004; 25: 438–441.
6) Dent Update. 2013; 40: 669–670.
7) Case Rep Rheumatol. 2015; 2015: 901795.
8) Case Rep Med 2017; 2017: 6327437
9) Lancet. 1983;2:121720.
10) Rheumatology. 2018;57:ii3ii10.
11) Otolaryngology—Head and Neck Medicine and Surgery. 2004;25, no. 6: 438–441.
12) N Engl J Med 2017;377:317-28.
13) A&R. 1990;33, no. 8:1122-1128.

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