Tongue Necrosis due to Giant Cell Arteritis

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ABSTRACT

Giant Cell Arteritis is a vasculitis that mainly affects women over 50. The most common manifestations are headache, jaw claudication, and amaurosis. If not diagnosed early, it can lead to rare irreversible ischemic consequences, with tongue necrosis being one of these. We report a case of a previously undiagnosed patient with lateral tongue necrosis who responded well to oral corticosteroid treatment. The diagnosis is clinical, laboratory and histological and may be aided by imaging exams. Initial treatment is with oral corticosteroids, with methotrexate and tocilizumab as alternatives. Diagnostic suspicion and quick start of treatment favorably influence the prognosis of the disease.

Keywords: Vasculitis, Giant cell arteritis, Tongue, Necrosis.

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INTRODUCTION

Giant cell arteritis (GCA) is a predominant vasculitis in women over 50 years. It involves medium and large caliber arteries, especially the carotid branches, especially the temporal arteries[1]. Headache, diplopia, amaurosis, and jaw claudication are common symptoms[2], and tongue necrosis is rare[3]. We report a case of GCA with oral corticosteroid-treated tongue necrosis and a good therapeutic response.

CASE REPORT

An 86-year-old female patient with headache developed right amaurosis, jaw claudication, and lingual burning. After seven days, she presented a necrotic plaque on the left side of the tongue. She had a history of Alzheimer's, systemic arterial hypertension, and diabetes mellitus.

On examination, she had a yellowish plaque on the right dorsal surface of the tongue and a necrotic plaque on the left lateral surface (Fig 1-A/B). We noticed the presence of pulse, pain, and thickening in the right temporal artery (Fig 2-A/B). Diagnostic investigation showed high ESR (82 mm/h) and other tests (electrocardiogram, cranial tomography, temporal artery, carotid and vertebral Doppler) without alterations. The diagnosis of complicated GCA with tongue necrosis was suggested.

Treatment was started with prednisone 40mg/day for 30 days with immediate symptom improvement. There was complete healing of the lingual lesion, although with tissue loss on the left lateral face of the tongue (Fig 3-A/B and Fig. 4) and persistence of amaurosis. After 7 months and slow and gradual weaning, the patient persists with the use of prednisone 2.5 mg every other day without pain symptoms or signs of recurrence.

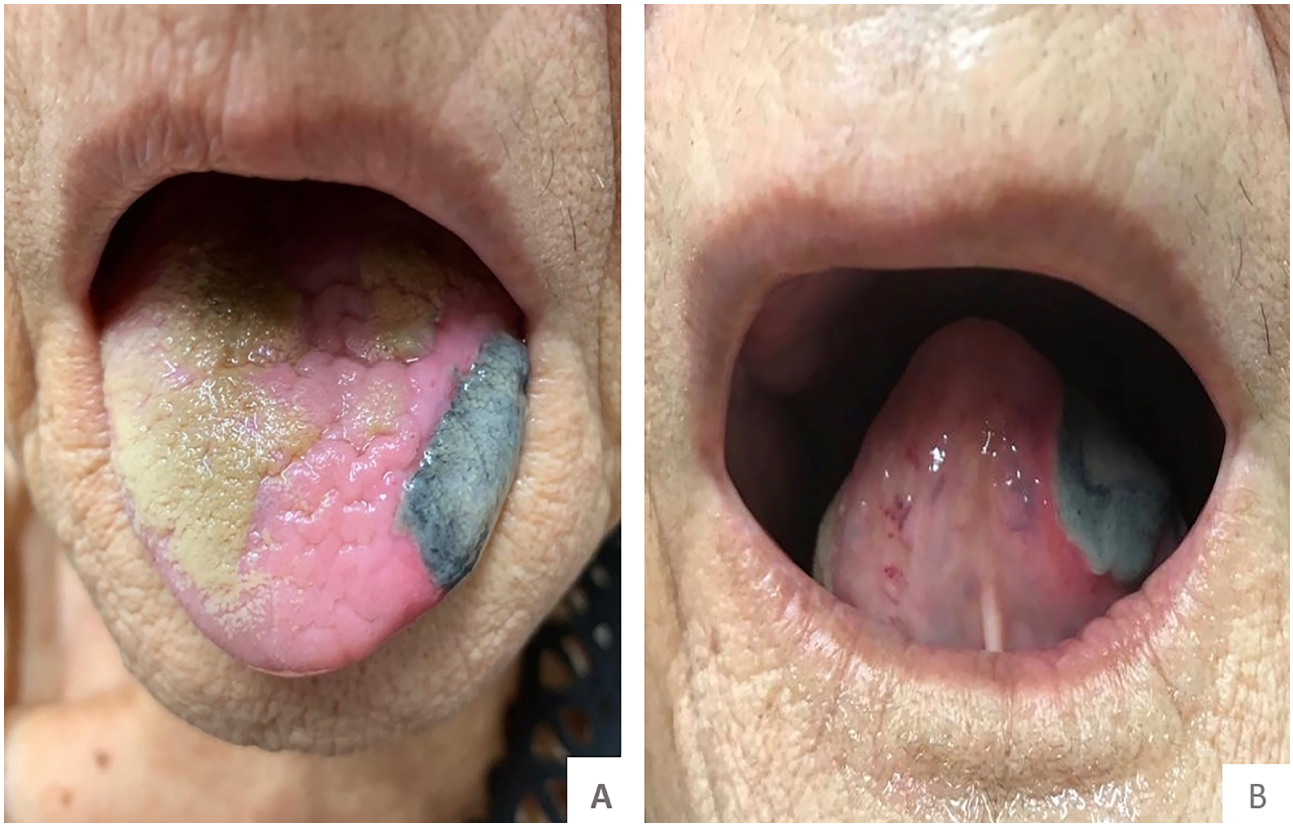

Figure 1. (A) Yellowish plaque on the right side of the tongue and necrotic plaque on the left side. **(B)** Necrosis extending from the ventral to the dorsal portion of the tongue.

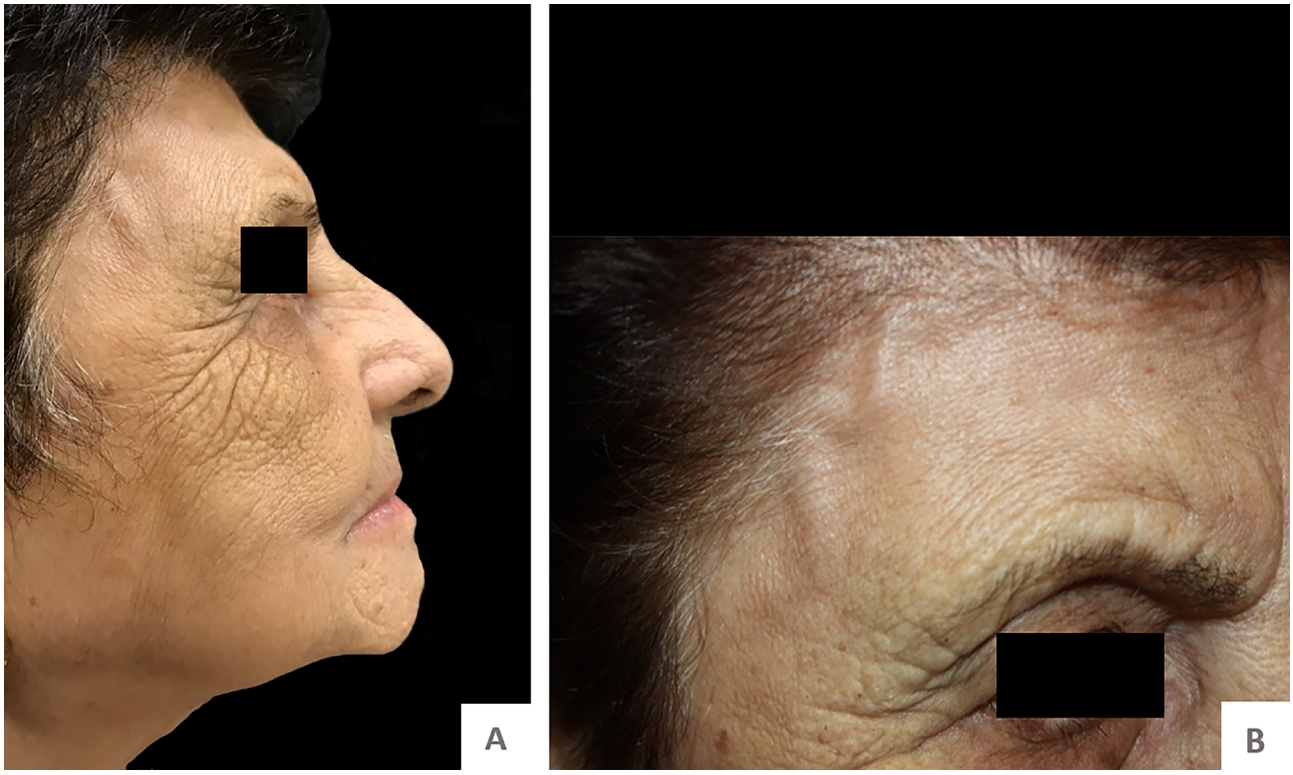

Figure 2. (A) and **(B)** Thick and tortuous right temporal artery.

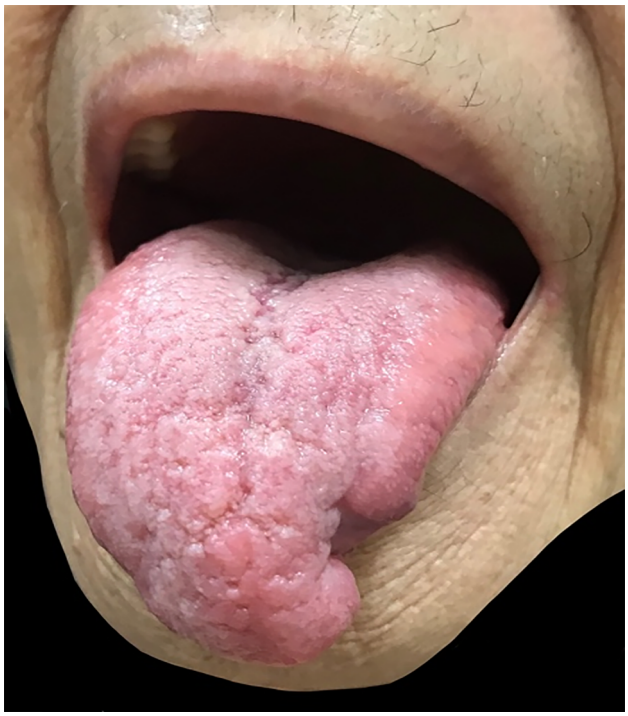

Figure 3. (A) and (B) Clinical response after 1 month of corticosteroid treatment.

DISCUSSION

GCA is a medium and large vessel vasculitis that especially affects the temporal artery. It is the most common form of primary systemic vasculitis, predominates in females at a ratio of 5:2 and occurs mainly in the elderly[4]. The clinical manifestations are diverse and vary according to the affected vessel, being common headache (90%), jaw claudication (50%), and amaurosis (40%). When there is tongue involvement, there may be edema, pain and lameness in 25% of cases. As the blood supply of the tongue is rich, necrosis of this tissue is infrequent and suggests a poor prognosis[1]. The high diagnostic suspicion indicates the need for early treatment in order to avoid serious complications such as stroke, cerebral artery dissection, and permanent amaurosis, as in the present case[5].

In the presence of tongue necrosis, other etiologies should be excluded such as embolism, carcinoma, radiotherapy, syphilis, and tuberculosis[1].

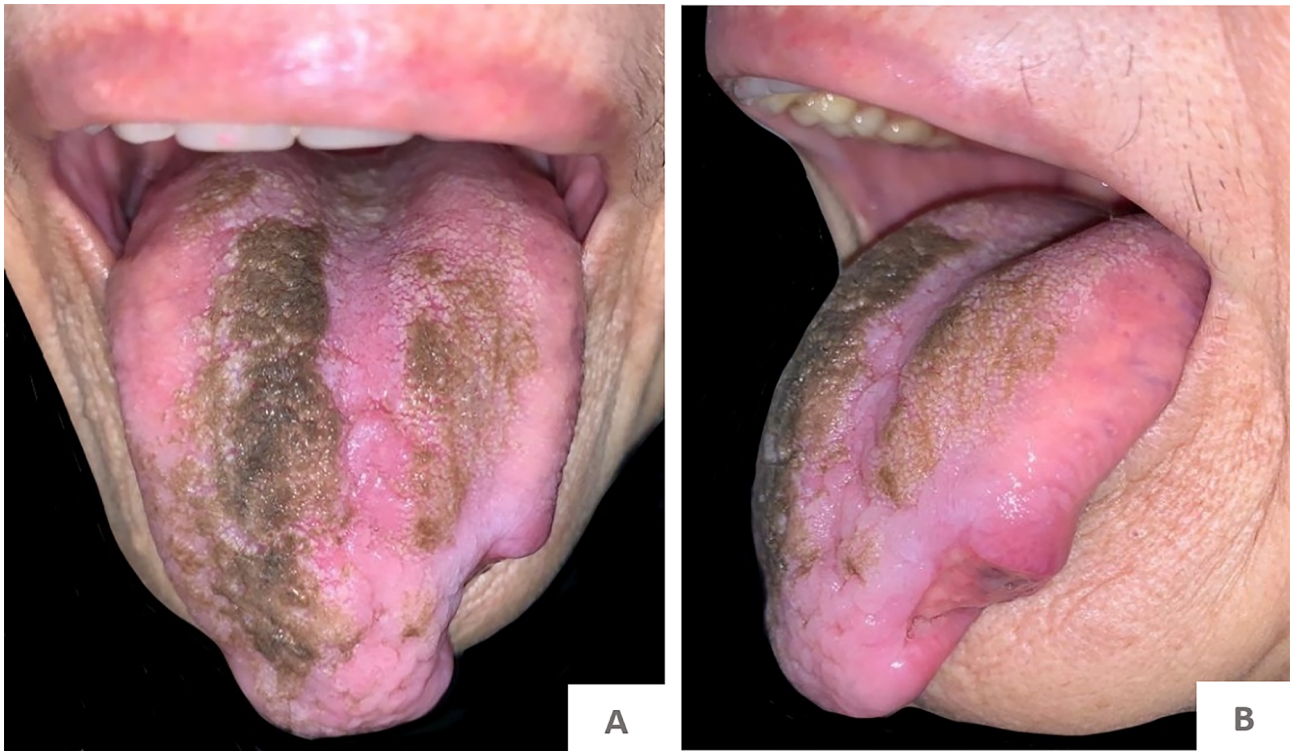

Figure 4. Clinical response after 2 months of corticosteroid treatment.

We discarded these possibilities and identified 4 of the 5 American College of Rheumatology criteria that confirmed the definitive diagnosis of GCA (Table 1) [6].

Temporal artery biopsy is the gold standard for diagnosis with a sensitivity of 54-92%. Doppler arterial ultrasound, whose specificity and sensitivity are 92.31% and 83.33%, respectively, may be useful[4]. In the reported case, there was no change in the latter, but the fact that it was performed after 8 weeks of treatment and was a dependent operator probably influenced the result.

Treatment is based on the use of corticosteroids at a dose of 1mg/kg/day for 4 to 6 weeks with subsequent weaning. Adjuvant therapy with methotrexate and tocilizumab may be instituted[7]. There is no consensus on the therapeutic period, but it is suggested to continue treatment for about 2 years to avoid relapse[8]. In this particular case, we were able to reduce prednisone in just 7 months without injury to the patient.

The clinical diagnosis of GCA is of fundamental importance and even the impossibility of performing a temporal artery biopsy should not be a reason to delay the initiation of treatment. Otherwise, it may lead to definitive squeals or more severe manifestations of the disease.

Table 1. Diagnostic criteria Giant Cell Arteritis according to American College of Rheumatology[7]

Criterion	Definition
1. Age at onset more than 50 yr	Development of symptoms or findings beginning aged 50 yr or older
2. New headache	New onset of, or new type of, localized pains in the head
3. Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to atherosclerosis
4. Increased ESR	ESR more than 50 mm/hr by Westergren method
5. Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear celll

GCA = giant cell arteritis; ESR = erythrocyte sedimentation rate; PMR = polymyalgia rheumatica

REFERÊNCIAS

1. DeBord LC, Chiu I, Liou NE. Delayed Diagnosis of Giant Cell Arteritis in the Setting of Isolated Lingual Necrosis. Clinical Medicine Insights: Case Reports2019; 12:1179547619857690. https://doi.org/10.1177/1179547619857690
2. Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. American journal of ophthalmology1998;125(4):509-20. https://doi.org/10.1016/s0002-9394(99)80192-5
3. Schurr C, Berthele A, Burghartz M, Kiefer J. Spontaneous bilateral necrosis of the tongue: a manifestation of giant cell arteritis? European archives of oto-rhino-laryngology2008;265(8):993-8. https://doi.org/10.1007/s00405-007-0556-x
4. Ball, E.L., Walsh, S.R., Tang, T.Y., Gohil, R. and Clarke, J.M.F. (2010), Role of ultrasonography in the diagnosis of temporal arteritis. Br J Surg, 97: 1765-1771. https://doi.org/10.1002/bjs.7252
5. Neto FXP, Carneiro KL, Junior OMR, Junior AGR, de Souza Jacob CC, Palheta ACP. Aspectos Clínicos da Arterite Temporal.2008;12(4):546-551.
6. Ling MLH, Yosar J, Lee BWH, Shah SA, Jiang IW, Finniss A, et al. The diagnosis and management of temporal arteritis. Clinical and Experimental Optometry2019. https://doi.org/10.1111/cxo.12975
7. Chean CS, Prior JA, Helliwell T, Belcher J, Mackie SL, Hider SL, et al. Characteristics of patients with giant cell arteritis who experience visual symptoms. Rheumatology international2019;39(10):1789-96. https://doi.org/10.1007/s00296-019-04422-5
8. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis & Rheumatism1990;33(8):1122-8. https://doi.org/10.1002/art.1780330810
9. Monti S, Águeda AF, Luqmani RA, Buttgereit F, Cid M, Dejaco C, et al. Systematic literature review informing the 2018 update of the EULAR recommendation for the management of large vessel vasculitis: focus on giant cell arteritis. RMD open2019;5(2):e001003. http://dx.doi.org/10.1136/rmdopen-2019-001003
10. Raza M, El Maideny Y, Bokhari N. Giant cell arteritis: advances in diagnosis and management. British Journal of Hospital Medicine2019;80(8):448-55. https://doi.org/10.12968/hmed.2019.80.8.448

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Conflicts of interest
None.

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