Tongue Necrosis due to Giant Cell Arteritis

Luiza Aguera Oliver, Alice Guiotti de Alencar, Marcelo Rosandiski Lyra, Ignacio Yáñez Silva

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.
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. Headache, diplopia, amaurosis, and jaw claudication are common symptoms, and tongue necrosis is rare. 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.
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. 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. 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.

In the presence of tongue necrosis, other etiologies should be excluded such as embolism, carcinoma, radiotherapy, syphilis, and tuberculosis.
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 useful⁴. 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 instituted⁷. There is no consensus on the therapeutic period, but it is suggested to continue treatment for about 2 years to avoid relapse⁶. 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 Rheumatology⁷

<table>
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<tr>
<th>Criterion</th>
<th>Definition</th>
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<tr>
<td>1. Age at onset more than 50 yr</td>
<td>Development of symptoms or findings beginning aged 50 yr or older</td>
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<tr>
<td>2. New headache</td>
<td>New onset of, or new type of, localized pains in the head</td>
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<td>3. Temporal artery abnormality</td>
<td>Temporal artery tenderness to palpation or decreased pulsation, unrelated to atherosclerosis</td>
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<td>4. Increased ESR</td>
<td>ESR more than 50 mm/hr by Westergren method</td>
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<td>5. Abnormal artery biopsy</td>
<td>Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell</td>
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GCA = giant cell arteritis; ESR = erythrocyte sedimentation rate; PMR = polymyalgia rheumatica
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Conflicts of interest
None.

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