To the Editor:

Giant cell arteritis or temporal arteritis (TA) is the most common systemic vasculitis in adults. Histologically there is a lymphocytic-monocytic panarteritis with the formation of granulomas and giant cells. There is patchy involvement of medium and large arteries, particularly the extracranial branches of the carotid artery and, more specifically, the temporal artery.

The clinical presentation of TA is variable, with multiple, nonspecific symptoms in elderly patients, particularly women, who initially present fever, asthenia, weight loss, headache, unpredictable joint and muscle pain, stiffness, and polymyalgia rheumatica. Up to a third of patients develop visual symptoms. Loss of vision, sometimes bilateral and irreversible, is the main complication of this condition and it can sometimes be the first symptom. The alterations in the blood tests in TA can also be relatively nonspecific, with normocytic-normochromic anemia, elevated alkaline phosphatase, and a raised erythrocyte sedimentation rate (ESR). Up to 40% of patients develop atypical clinical manifestations such as respiratory symptoms, pyrexia of unknown origin, aortic aneurysms, or digestive tract, neurological, or skin disorders.

We present the case of a 91-year-old woman with a past history of systemic hypertension, congestive cardiac failure, and atrioventricular block for which a pacemaker had been implanted. She came to the emergency department with a 48-hour history of bilateral loss of vision, with no accompanying symptoms.

On physical examination there was asymmetric, unreactive, bilateral mydriasis with changes suggestive of ischemic optic neuritis on funduscopic examination. The blood test performed in the emergency department revealed a normocytic anemia (hemoglobin, 10.7 g/dL) and an ESR of 23 mm/h. Cerebral computed tomography showed no relevant abnormalities. On a suspicion of TA, biopsy of the temporal artery was performed and treatment was started with intravenous methylprednisolone at a dose of 250 mg every 6 hours.

Histological study of the artery showed a thickened wall with a moderate chronic inflammatory infiltrate in the media and adventitia of the vessel, composed of lymphocytes, histiocytes, occasional eosinophils, and occasional images suggestive of giant cells (Figure 1).

Two days after starting the treatment, the patient presented intense pain in the tongue, leading to difficulty moving the tongue, swallowing, and speaking. On examination, there was a large, well-defined, deep, excavated ulcer with a clean base and that was not infiltrated on palpation (Figure 2); it was very painful.

On a repeat blood test, the ESR had risen to 88 mm/h, with no other abnormalities.

Biopsy of the tongue lesion showed a deep ulcer that reached the skeletal muscle tissue, with fibrosis and images of myocyte necrosis, suggestive of ischemia (Figure 3).

Figure 1. Temporal artery with a thickened wall and a moderate inflammatory infiltrate in the media and adventitia. Hematoxylin-eosin x4.

Figure 2. Very painful ulcer on the right side of the tongue; the lesion was not infiltrated on palpation.
Intravenous, high-dose corticosteroid treatment and analgesia were maintained, and there was a progressive reduction in the size of the ulcer on the tongue, leading to complete re-epithelialization. However, the loss of vision persisted.

The diagnosis of TA can sometimes be difficult as the early symptoms are nonspecific. Age is the principal risk factor; the disease is virtually unknown below the age of 50 years, and the incidence then gradually increases—1.5 per 100,000 population in the sixth decade of life, 20.7 per 100,000 population in the eighth decade. The most common manifestation is unilateral headache, particularly affecting the temporal region. The abnormalities of the temporal artery, such as thickening, pain on palpation, or a reduction or absence of the pulse not justified by other causes such as arteriosclerosis, are considered diagnostic criteria. The ESR is almost always raised in TA, and can reach over 100 mm in some cases. However, the ESR is below 50 mm in up to 10% of patients, and even below 30 mm in 4% of cases, as occurred in our patient. Biopsy of the temporal artery usually confirms the diagnosis, although false negatives occur in up to 5%-10% of cases due to the patchy nature of the histological changes, making a second biopsy or bilateral biopsies necessary. The inflammatory changes in the artery can persist for weeks after starting treatment. Dermatological abnormalities are uncommon in TA. Necrosis of the scalp is the most commonly reported cutaneous sign. It appears that this could be a marker of severity, and it occurs in a subgroup of older patients with a higher incidence of loss of vision and a mortality of up to 40%. Still less common is involvement of the tongue, in the form of pain, stiffness, ulceration, or extensive necrosis.

When the diagnosis is suspected, TA must be treated rapidly to avoid irreversible complications, particularly complete loss of vision. TA usually responds well to high doses of corticosteroids. The ischemic disorders, including those of the skin, can appear at any time in the course of the disease, particularly during the tapering of steroid treatment; these patients must therefore remain on treatment for long periods, sometimes for years or even for life.

TA must be included in the differential diagnosis of tongue disorders in elderly patients with heterogeneous clinical presentations with multiple, nonspecific symptoms with no other apparent cause.

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Conflicts of interest
The authors declare no conflict of interest

References