The Sport's Bar Grandpa: an unusual left temporo-mandibular and tongue pain

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ABSTRACT

This case report describes the condition of an elderly patient admitted to hospital for a new onset headache and pain in the left temporo-mandibular joint, initially incorrectly interpreted as an angioedema, but that evolved into a tongue infarction.

Case Report

Mr. BE, male, 78-year-old, a former television actor in a fiction where he played the role of the grandfather at the Sport's Bar. For one month he had pain in the temporo-mandibular joint with an ipsilateral temporal headache, combined with difficulty in chewing, swallowing and sore throat. He was examined by an ear-nose-throat (ENT) specialist, who initially attributed the pain to a possible Costen syndrome or a jaw malocclusion. A few days after, following an insect bite on the right thigh treated with azithromycin, he was admitted to hospital with a diagnosis of lingual *angioedema*, owing to reported tongue bloating and respiratory distress, with severe headache and pain in the left side of the tongue. The physical examination on admission showed the lack of the temporal left pulse and a reduction in the right-side pulse. ENT examination was essentially negative, except for the presence of severe pain on palpation on the left ventral surface of the tongue, and a few small whitish areas in the oral cavity, interpreted as initial candidiasis; tongue edema was not detectable. Laboratory tests (main data) showed neutrophilic leukocytosis, mild normocytic anemia, elevated erythrocyte sedimentation rate (ESR) (53 mm/h) and an increase of C-reactive protein (CRP), hyper-α_2-globulinemia, with hypoalbuminemia and albumin to globulin (A/G) ratio inversion, mild fasting hyperglycemia, a modest increase in serum creatinine, negative antinuclear antibodies and anti-neutrophyl cytoplasmatic antibodies. An ENT control carried out four days following the first visit highlighted a large well-demarcated bluish area, where the patient complained of pain in the tongue, covered with fibrin for epithelial disintegration, which was assumed to be a potential acute lingual ischemia (Figure 1).

Our patient presented a new onset headache with left temporo-mandibular pain, left mandibular dysfunction with lingual pain, sore throat, absence of left temporal pulse, signs of left tongue ischemia, elevated ESR and CRP. A potential left tongue infarction was suspected and an oral and maxillofacial computed tomography scan was performed: the left tongue appeared relatively hypodense with absence of the left lingual artery, despite the contrast media. A maxillo-facial angiographic examination by trans-femoral catheterization and selective injection of the left external carotid arteries was performed: the right lingual artery appeared to be thread-like, the left lingual artery appeared to be occluded at its origin with no signs of any significant effective collateral circulation (Figure 2).

An echocolordoppler of the epiaortic vessels showed a fibro-calcific atheromasia of the common carotid and of the bulb origin of the left internal carotid artery, in the absence of hemodynamic stenosis of cerebral afferent vessels bilaterally, with vertebral patency. Despite the absence of bioptic data, a giant-cell arteritis with tongue infarct was suspected. Corticosteroid therapy was undertaken with 80 mg/day metilprednisonolone. After three days without significant clinical response, in particular for the headache, the steroid was administered as

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a bolus injection of 1 g intravenously per day, in a single morning dose for three days, which led to a prompt and excellent clinical response, with the disappearance of the headache and the temporo-mandibular joint pain. The pain in the left tongue still persisted and was controlled with oral morphine sulphate at a dose of 10 mg 4 times daily. Methylprednisolone was administered at a dose of 120 mg per day with subsequent very slow tapering in the order of 10-15% every week. Along with this treatment, also acetylsalicylic acid 100 mg/day and enoxaparin at the dose of 6000 UI were administered twice daily. Since the patient could not eat due to the oropharyngeal pain, he was first fed by total parenteral nutrition and, subsequently perendoscopic gastrostomy (PEG). A naso-gastric tube was excluded, because of the persisting pain. During the hospital stay there were two episodes of respiratory distress due to inhalation, the latter during PEG feeding. After the medical treatment, the headache disappeared and the inflammatory markers were significantly reduced.

Subsequently, the patient gradually showed signs of recovery during refeeding, with progressive general improvement as well as trophic lesions of the tongue (Figure 3).

Discussion

Giant-cell (GCA)-Horton's temporal arteritis is an inflammatory disease that typically affect adults, mostly in white individuals older than 50 years of age and women are affected ~2-3 times more often than.[1-3] Symptoms may be different and unusual, depending on the affected artery. The typical clinical manifestations of GCA are new headache, jaw claudication and visual loss,[4] nevertheless approximately 40% of patients with GCA show initially an atypical picture. Clinical manifestations of the disease include systemic symptoms, such as fever and weight loss, malaise, depression, headache for involvement of the temporal or occipital arteries, mandibular dysfunction with difficulty to chew hard food, discomfort or pain in the jaw, dental pain, tongue pain, widespread myalgia (polymyalgia rheumatica), visual symptoms with partially obscured vision, amaurosis fugax, diplopia, visual field changes up to sensory loss due to posterior ciliary artery occlusion (Figure 4).[5-9]

Less common and rare findings are: sore tongue and/or throat, uneven pulses or uneven blood pressure in the upper limbs, aortic regurgitation murmur, mononeuritis multiplex, dysarthria, hearing loss, mesenteric ischemia. Also the necrosis of the scalp and an ischemic involvement of both lingual and occipital arteries have been also described. In our case the left lingual artery was completely occluded and involved and caused an infarction of the left side of the tongue. The relatively early detection of this clinical condition has allowed us to achieve more than satisfactory results in improving initial symptoms. The diagnosis was made only on the basis of clinical and instrumental elements, even if temporal artery biopsy, represents the gold standard for diagnosing GCA.[6] It should be pointed out, however, that the clinical assessment is very important. The American College of Rheumatology criteria for the diagnosis of GCA include, besides a positive temporal artery biopsy, an age over 50 years, a new appearance of any type of head pain, abnor-

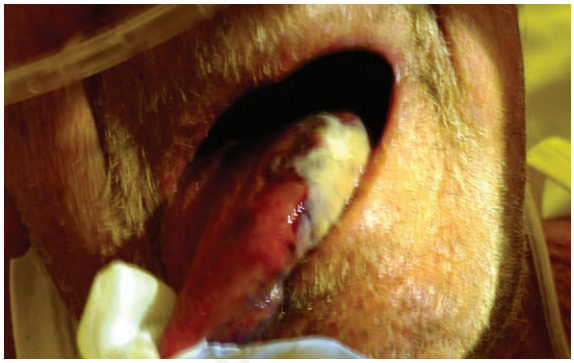

Figure 1. Infarction of the left side of the tongue.

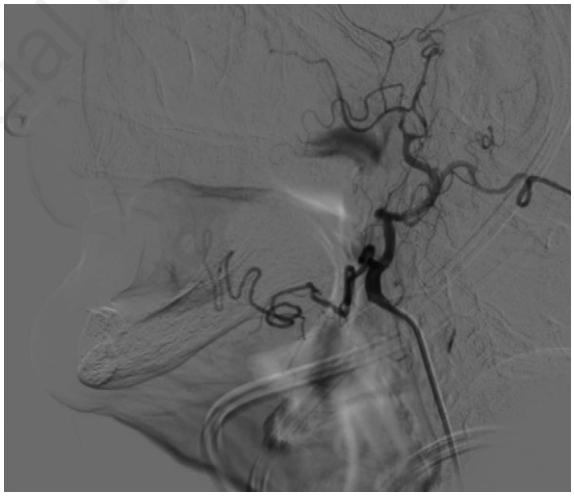

Figure 2. Left lingual artery amputated at the beginning.

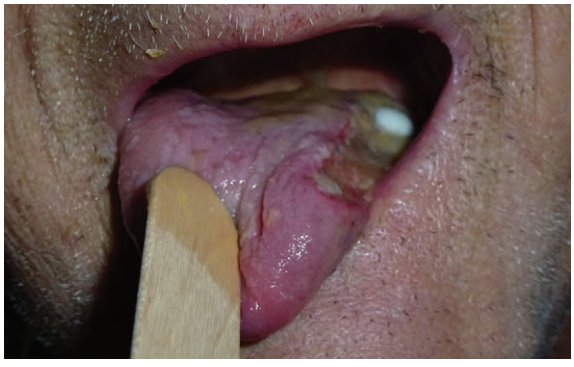

Figure 3. Improvement of trophic lesions of the tongue.

mal accentuation, prominence, tenderness of temporal artery, ESR ≥50 mm/h (Figure 5).[2]

To confirm the diagnosis at least 3 out of 5 criteria have to be present: with 3 or more of these findings the sensitivity and the specificity of the evaluation method are respectively 93.5% and 91.2%.[2] The presence of certain symptoms increases the probability [likelihood ratio (LR)] of disease: in particular, the mandibular dysfunction involves a positive LR of 4.2, diplopia of 3.4, the accentuation, prominence or tenderness of the temporal artery, of 4.6, 4.3 and 2.6 respectively[10,11] (Figure 6).

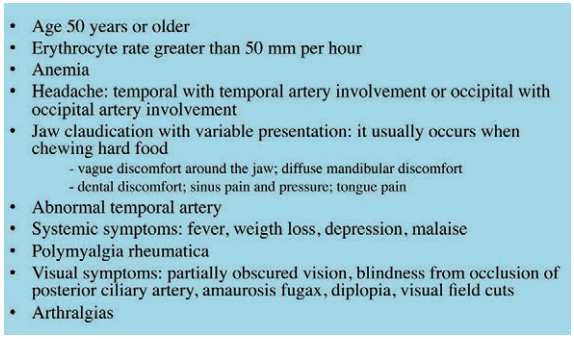

Figure 4. Typical features associated with giant-cell arteritis.

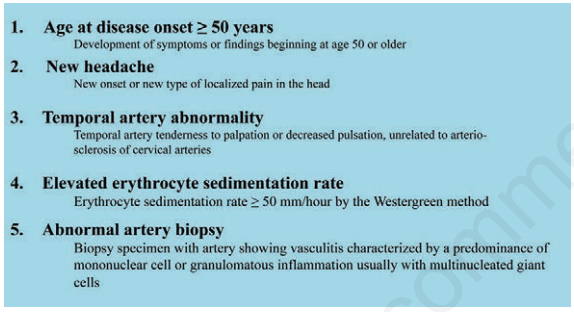

Figure 5. The American College of Rheumatology criteria for the diagnosis of giant-cell-Horton's temporal arteritis. For classification purposes, giant-cell (temporal) arteritis is confirmed when at least 3 of these 5 criteria are present. The presence of any 3 or more criteria yelds a sensitivity of 93.5% and a specificity of 91.2%.

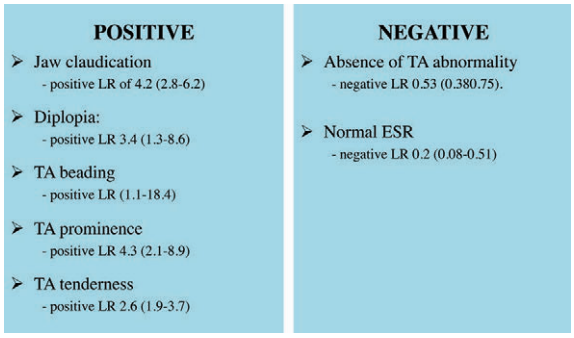

Figure 6. Symptoms which increase the probability of disease. LR, likelihood ratio; TA, temporal artery; ESR, erythrocyte sedimentation rate.

Conclusions

Given the broad spectrum of clinical manifestations of GCA, it is necessary for physicians to consider also an unusual and atypical presentation[12] of the disease in order to reach an early diagnosis. One of these may be tongue infarction,[13-15] which, together with scalp necrosis, is regarded in international literature as one of the rarest forms of the potential early manifestations of GCA.

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