Tongue ischemia – an unusual presentation of giant cell arteritis

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ABSTRACT

Unusual presentations of Giant Cell Arteritis (GCA) can sometimes delay the diagnosis and its prompt treatment. An 83-year-old male patient was admitted in the emergency department with a few hours evolution of tongue swelling, dysphagia and dysarthria. He also complained of a bitemporal headache with about 4 months of evolution and resistant to all treatment prescribed, including ergotamine, that he had started one week before. Upon examination, the patient presented a bilateral temporal pain and reduced mobility of the tongue which evolved to complete cyanosis. The blood tests revealed normocytic normochromic anaemia, an erythrocyte sedimentation rate of 62 mm/h, and C--reactive protein of 23,6 mg/dl. Cranial CT scan was normal and the cervical CT angiogram showed reduced vascularization of the left submandibular gland and of the base of the tongue. The cervical doppler ultrasound was compatible with arterial inflammation. Given the high suspicion of GCA, the patient was immediately put on a high dose of corticosteroid, resulting in a big improvement of the symptoms, which continued in the following weeks.

In conclusion, the clinical suspicion of GCA is fundamental for an early diagnosis. The authors consider that ergotamine might have triggered tongue ischemia in this case.

Keywords: Giant cell arteritis; Vasculitis; Tongue necrosis; Ergotamine.

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INTRODUCTION

Giant cell arteritis (GCA) is a medium-large vessel vasculitis that predominantly affects the extracranial branches of the carotid arteries. Its diagnosis is obtained through clinical and laboratory findings. According to the American College of Rheumatology, the presence of at least 3 of the following criteria is highly suggestive of ACR: aged 50 or more, a new headache, temporal artery abnormality, an erythrocyte sedimentation rate (ESR) of 50mm/h or more, and abnormal artery biopsy[1]. Unusual presentations can sometimes delay the diagnosis and the prompt treatment, which is indispensable for a good prognosis[2]. As the tongue has an huge collateral blood supply and numerous anastomoses, an extensive vascular involvement is needed for lingual manifestations[3]. Thus, tongue ischemia is a rare initial manifestation of GCA, appearing in less than 1% of the cases[4].

CLINICAL CASE

An 83-year-old male patient, with a medical history of asthma, dementia and prostate carcinoma, was admitted to the emergency department with a few hours evolution of tongue swelling, dysphagia and dysarthria. The patient also complained of a bitemporal headache with about 4 months of evolution and resistant to all treatment prescribed. One week before, he was started on ergotamine with no improvement. He denied other symptoms. Upon examination, the patient presented bilateral temporal pain and reduced mobility of the tongue that evolved to complete tongue cyanosis within a few hours (Figure 1-A). The performed blood tests revealed normocytic normochromic anaemia (haemoglobin of 11,6 g/dl), an ESR of 62 mm/h, and C-reactive protein of 23,6 mg/dl. Cranial CT scan was normal, and the cervical CT angiogram showed reduced vascularization of the left submandibular gland

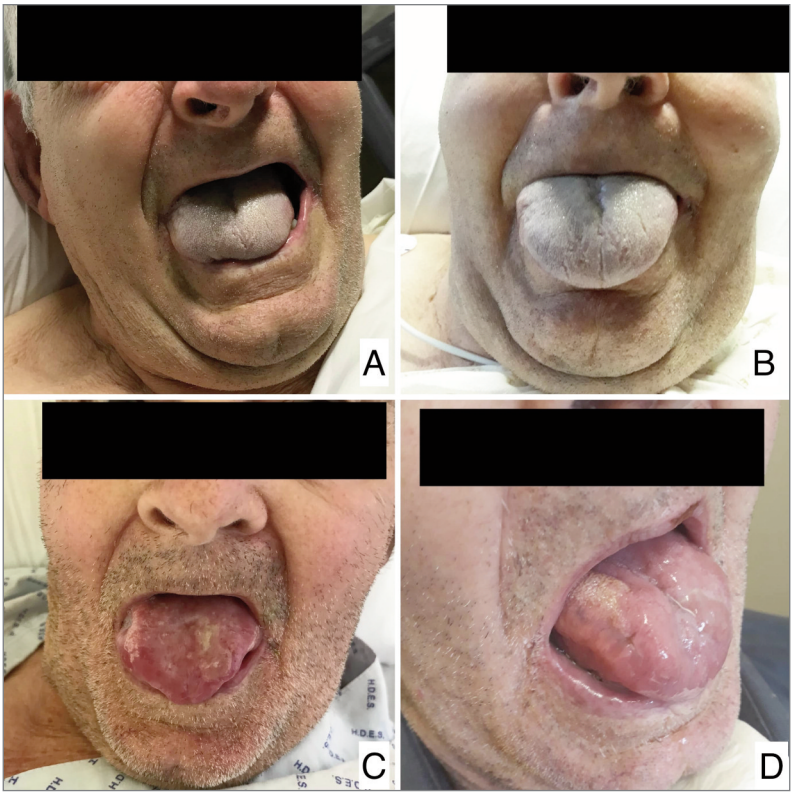

FIGURE 1. Evolution of the tongue's coloration and mobility throughout the treatment. A presents the patient at admission; B, C and D portray, respectively, the patient 1, 2 and 3 weeks after the initiation of corticotherapy.

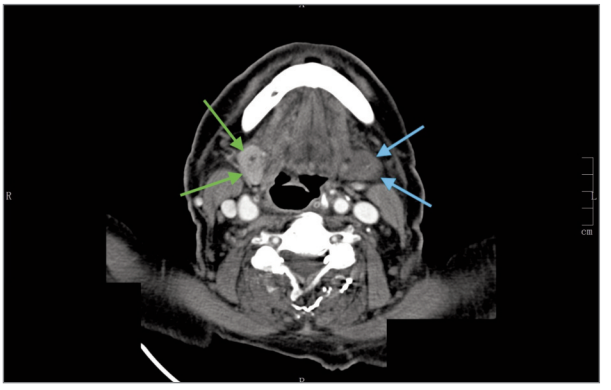

FIGURE 2. CT angiogram showing reduced vascularization of the left submandibular gland (blue arrows). The green arrows show the normal vascularization of the right submandibular gland.

and of the base of the tongue (Figure 2). The cervical doppler ultrasound was compatible with arterial inflammation. Given the high suspicion of GCA, the patient was immediately started on a high dose corticosteroid (1g methylprednisolone for three days, followed by prednisolone 1 mg/kg). The temporal artery biopsy, done on the third day of the corticotherapy, showed no evidence of GCA. One week later, there was an improvement of the tongue discoloration (Figure 1-B) and of its mobility, which continued in the following weeks (Figure 1-C). The corticosteroid was progressively tapered, and three months later there was complete resolution of the symptoms (Figure 1-D) and improvement of the laboratorial parameters (haemoglobin of 13,1 g/dl; ESR of 15mm/h and C-reactive protein of 0,190mg/dl).

DISCUSSION

We should suspect of tongue ischemia in the presence of tongue claudication, blanching, glossitis, loss of taste, pain, oedema, cyanosis and erythema[4]. The authors consider that ergotamine might have triggered tongue ischemia in this particular patient. Although temporal artery biopsy is the gold standard test for GCA diagnosis, around 30% of the cases can have the

disease even with a negative biopsy, as the sensibility ranges between 70-90%[5]. Thus, the clinical suspicion should prevail, and the corticotherapy should be started as soon as possible to prevent damages.

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