Catastrophic Presentation of Giant Cell Arteritis

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Introduction: Giant cell arteritis (GCA) is a chronic inflammatory vasculitis that predominantly affects large- and medium-sized arteries in individuals over 50 years old with heterogeneous presentation. Objectives: describe a case with atypical and catastrophic presentation.

Materials and Methods: A 76-year-old woman presented with a sudden onset of right hemitongue stroke, followed by the development of progressive bilateral blindness, more severe in the right eye, and mandibular claudication one month later. Neurological examination was normal. However, general physical examination revealed the presence of hardened bilateral temporal arteries to palpation. Ocular examination showed complete blindness in the right eye and uncertain light perception in the left, with bilateral papillary edema. Facial computed tomography revealed hypodensity in the right lingual area, with a dominant left artery. A temporal artery biopsy demonstrated giant multinucleated cells, consistent with a diagnosis of GCA. Additionally, a tongue biopsy revealed extensive ischemic necrosis with relative sparing of the muscle layers, which were involved by a dense eosinophilic granulocytic inflammatory infiltrate. Laboratory results showed an elevated erythrocyte sedimentation rate (ESR) of 100 mm/h (normal: 1-32 mm/h), C-reactive protein (CRP) of 56.5 mg/L (normal: <5 mg/L). Other immunological exams were unremarkable, and the patient's immunological history was negative. Her medical history included polyosteoarthrosis, atrioventricular block Mobitz II, diabetes (treated with metformin), and arterial hypertension. Brain MRI and angio-CT were normal. A PET scan for staging revealed mild uptake in the femoral and popliteal arteries.

Results: The patient was diagnosed with GCA, a systemic inflammatory condition. During hospitalization, she underwent pacemaker insertion due to complete heart block (third-degree AV block, Mobitz II). She was treated with intravenous Tocilizumab therapy (8 mg/kg).

Discussion: The tongue's primary blood supply comes from the lingual artery, a branch of the external carotid artery. In GCA, inflammation can lead to reduced blood flow, resulting in ischemia and necrosis of the tongue tissue. This is usually unilateral but may sometimes involve the entire muscle. Patients may present with symptoms such as lingual pain, ulcers, edema, pallor, and, in severe cases, necrosis of the tongue tissue. To date, fewer than 30 cases of tongue necrosis in the context of GCA have been reported in the literature, with some cases involving bilateral tongue necrosis, though this is rare. Conclusion: The clinical presentation, imaging, and laboratory findings underscore the importance of considering GCA in elderly patients with unexplained vascular symptoms, such as lingual necrosis.

References:

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