CASE DESCRIPTION

A 51-years-old man came to the emergency room of our department for impairment in tongue mobility and speech, which started two days before. The patient was asymptomatic, except for reporting a “bulky tongue” sensation and an extremely mild hemicrania, localized to the right temporal area. Anamnesis was negative for pathologies and for drug intake (blood pressure 125/74 mmHg). No history of head or cervical trauma was declared, as no previous loco-regional infections was reported. At the extra-oral examination, no cervical lymphadenopathy or submandibular mass could be observed. At the intra-oral examination, tongue sensitivity and taste perception were normal, no hoarseness or change in the voice were evident, nor referred by the patient himself. No signs of dysphagia could be found. Tongue mobility was significantly reduced, in absence of mucosal lesions, together with the loss of the right tongue’s muscular tone, detectable at palpation. The right tongue appeared enlarged when lying in the floor of the mouth, while right deviation was observed during tongue protrusion (Fig.1). There was no evidence of Horner’s syndrome or other focal neurological deficits.

Intra-oral clinical picture, thus, excluded lesions involving the oral epithelium. A contrast-enhanced magnetic resonance imaging (MRI) of the complete head and neck area (neck, maxillofacial region, skull and brain) and subsequent CT angiography were performed (Fig.2).

What is your diagnosis?

RESPONSE

Brain MRI was normal and no suspicious masses were observed in the maxillofacial region, excluding submucosal neoplastic lesions of the tongue. MRI of the neck revealed intramural hematoma of the right carotid artery assuming compression of the right hypoglossal nerve (Fig.2). The subsequent neck CT angiography confirmed the diagnosis of extracranial right internal carotid artery dissection (ICAD). According to Cervical Artery Dissection In Stroke Study (CADISS), antiplatelet therapy with acetylsalicylic acid was started [1]. At 6 weeks follow-up the tongue mobility was normal, headache and mild dysarthria were completely regressed.

During ICAD, a tear occurs in the tunica intima of the artery and the blood, entering the arterial wall under pressure, separates the wall layers and causes the formation of a false lumen. This process results in the formation of an intramural hematoma which may potentially occludes the vessel. ICAD could lead to cerebral thrombo-embolism and/or hypoperfusion, with subsequent ischemic stroke. Artery dissection may also result in the compression of surrounding structures causing cranial nerve palsies or Horner's syndrome. ICAD can causes hypoglossal nerve paralysis through direct compression of the expanded dissected arterial wall or through vascular interruption of the vasa nervorum, which feed the nerve [2,3]. In the most cases, ICAD affects the extracranial segment, while the intracranial part may be rarely involved [3]. ICAD is an important cause of ischemic stroke in young adults, accounting for 25% of all cases in subjects under 45 years, and it affects both genders with an annual incidence of 3 cases per 100,000 [2,3]. Most cases occur spontaneously or can be associated to minor (neck manipulation, sneezing, coughing) or major (road traffic accidents) neck traumas. An additional etiological role has been suggested for infections of the upper respiratory tract, connective tissue disorders, arterial hypertension, fibromuscular dysplasia, osteogenesis imperfecta and estrogen-progesterone therapies [3]. ICAD typically presents with headache, ipsilateral cervicofacial pain, Horner's syndrome, tinnitus, transient or complete cerebral ischemic symptoms [3]. Only 0.5% of the patients affected by ICAD

are painless, while approximately 95% of them reports pain as main symptom at onset [4]. Moreover, ICAD may be associated to cranial nerve palsy in 12% of the cases and the hypoglossal nerve is the most frequently involved in 5% of the patients [2,3]. The multicenter randomized study CADISS did not show any differences between the efficacy of antiplatelet and anticoagulant therapies in cervical artery dissection; in case of an extracranial ICAD, it is therefore recommended to use any of these therapies [1,2]. The treatment duration is mainly related to the results of imaging performed during follow-up and the prognosis is generally good in more than 90% of cases [2]. Isolated hypoglossal nerve palsy in painless patient without any other findings, as presented in our case, is a rare clinical manifestation of ICAD.

Hypoglossal nerve palsy may be related to various pathological processes. Differential diagnosis should include: submucosal tongue malignancies, primary skull base neoplasms (e.g., paragangliomas), bony skull base metastases, traumatic skull base fractures and internal carotid artery dissection [4,5]. The main symptoms associated to hypoglossal nerve palsy include dysarthria, altered swallowing and impaired tongue movement [3]. Hypoglossal nerve palsy is a rare complication of ICAD, but in some cases it may be the only clinical manifestation of this disease [3]. Magnetic resonance imaging of the head and neck and CT-angiography of the neck vessels, should be considered if ICAD is suspected; brain CT is often performed in patients with suspected stroke, but it does not allow to highlight ICAD [2,3].

ICAD is just one among many disorders which can effort the hypoglossal nerve along its anatomical course. However, considering that ICAD is a treatable cause of ischemic stroke in young adults, it is mandatory to include it in differential diagnosis of hypoglossal palsy.

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Figure legend

Fig.1: Right tongue deviation on protrusion in absence of other clinical signs and symptoms.

Fig.2: MRI of the neck was suggestive of extracranial right internal carotid dissection.

Disclosure of interest

The authors have no relevant financial or non-financial interests to disclose.

Funding

The authors received no specific funding for this work. No funds, grants, or other support were received.

Patient's informed consent

Data and samples were collected under patient's informed written consent, guaranteeing anonymity.

Contributors section

N.L. wrote the manuscript, collected the clinical data and followed the patient during diagnosis; A.S. reviewed the manuscript and followed the clinical case; G.L. reviewed the manuscript and followed the patient during diagnosis.