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Isolated hypoglossal nerve palsy due to internal carotid artery dissection

R. Shahab, M.D., F.R.C.S., L. E. Savy, BSc, M.B. B.S., F.R.C.R., C. B. Croft, F.R.C.S., F.R.C.S. (Ed.), T. Hung, F.R.C.S.

Abstract
A case of an isolated hypoglossal nerve palsy is reported. The differential diagnosis is discussed, in the context of the requirement for careful scrutiny of the entire course of the hypoglossal nerve on imaging, to detect underlying pathology remote from the tongue, and to avoid unnecessary invasive diagnostic procedures prompted by the appearance of a 'pseudomass' of the weak tongue both clinically and radiologically.

Key words: Hypoglossal Nerve; Paralysis; Carotid Artery, Internal, Arteriosclerosis

Case report

A 46-year-old man presented with a short history of difficulty swallowing and slurred speech, which he attributed to difficulty moving his tongue. Two weeks earlier he had had a sore throat and had been prescribed antibiotics. He attended our institution, having had computed tomography (CT) and magnetic resonance imaging (MRI) of the head and neck in his own country, which had been reported as showing swelling of the left tongue base but no other abnormality.

There was no significant past medical history and no history of any head or neck trauma or previous operations. The patient smoked about 25 cigarettes a day and drank alcohol occasionally. Examinations revealed a left-sided tongue weakness with fasciculation of the left lateral border of the tongue. The remaining cranial nerves were

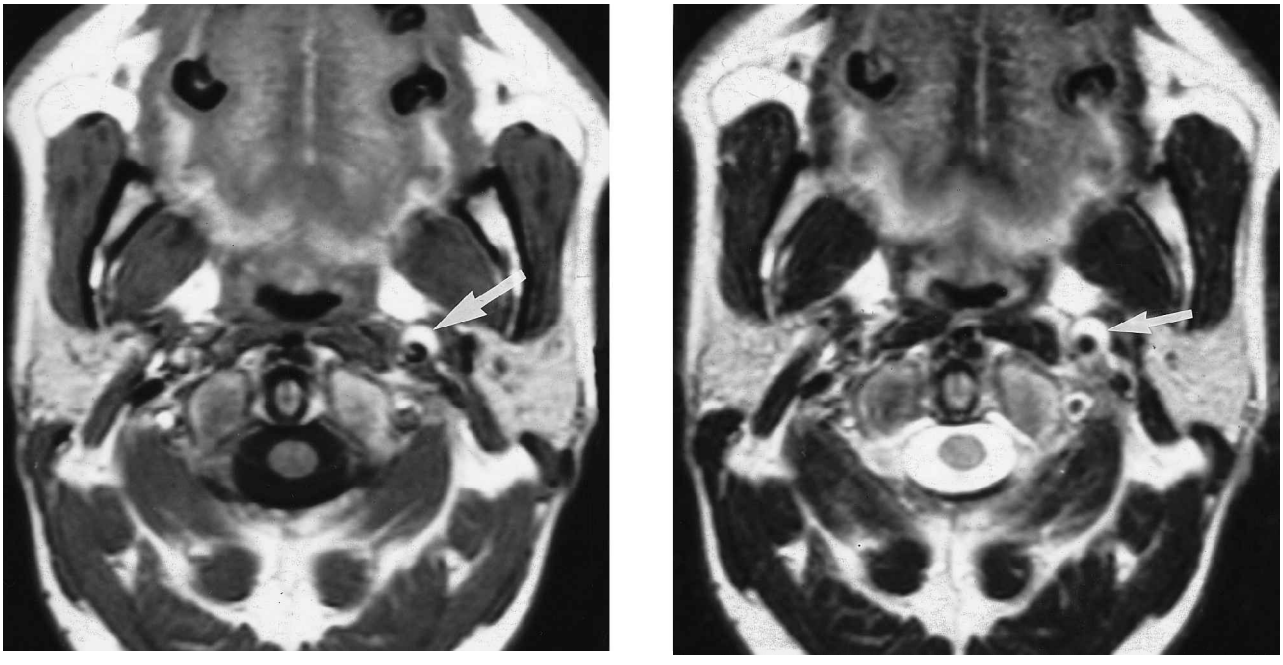

Fig. 1

(a) T1-weighted and (b) T2-weighted axial MRI, both showing crescentic high signal (arrow) due to intramural haematoma around the low signal (flow void) of the patent left internal carotid artery lumen. The dissection of the vessel wall occupies the space lateral to the artery in the position of the hypoglossal nerve. Also note the asymmetry of the tongue base, bulkier on the left side, but without any symmetry of signal on either T1-weighted or T2-weighted images.

From the Royal National Throat Nose & Ear Hospital, Gray's Inn Road, London, UK
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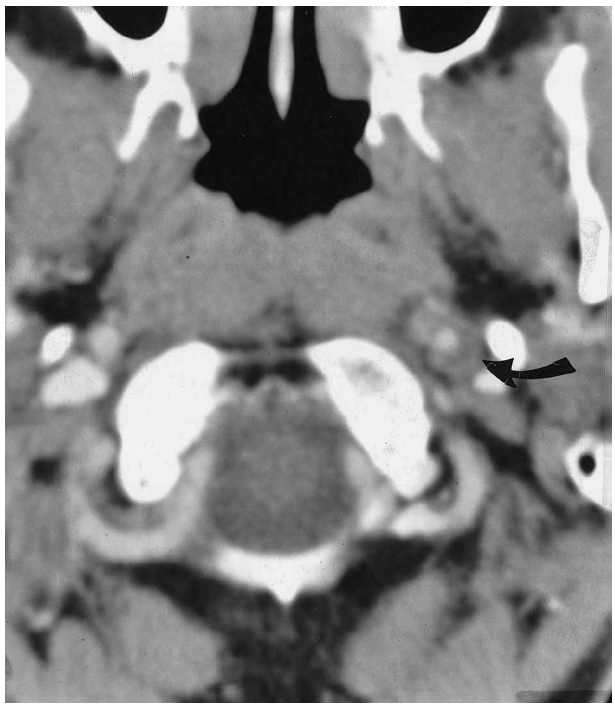

FIG. 2

Contrast-enhanced CT showing slightly reduced calibre of the enhancing left internal carotid artery lumen with a cuff of non-enhancing low density around it due to intramural haematoma and a peripheral dense rim corresponding to the adventitia (arrow).

intact. The left tongue base was prominent on mirror and fibreoptic examination but on palpation no abnormality could be detected. The rest of the ENT examination was unremarkable. Full blood count, erythrocyte sedimentation rate, and angiotensin-converting enzyme were all normal. A diagnosis of left peripheral hypoglossal nerve palsy was made and, because no obvious cause could be detected, the patient underwent a panendoscopy, examination under anaesthesia (EUA), and biopsy of the left nasopharynx, tonsil and tongue base. The panendoscopy, EUA and biopsies did not show any abnormality.

The CT and MRI scans were reviewed by a neuroradiologist. There was asymmetry of the tongue base which was bulkier on the left. There was no pathological signal or density of the tongue base itself, nor within the intrinsic muscles of the tongue. On all axial MRIs (T1-weighted, T2-weighted and FLAIR), there was semilunar, crescentic, high signal around the upper cervical segment of the left internal carotid artery in a spiral configuration anterior, lateral and posterior to the vessel, seen from the level of the C2 vertebra up to the proximal part of the carotid canal (Figure 1). The lumen of the artery did not appear markedly narrowed. The intracranial segments of the internal carotid appeared normal, and there was no abnormality of the brain parenchyma. Contrast-enhanced CT showed a cuff of low density around the enhancing arterial lumen at the same site, with a dense, well defined rim (Figure 2). On CT the arterial lumen did appear narrower than that of its fellow on the right. The advice of a vascular surgeon was taken. A repair of the aneurysm was not considered necessary. The patient was treated with aspirin (75 mg once daily) as several weeks had elapsed without progression of the neurological deficit. A six-month follow-up appointment was made to monitor the 12th nerve function.

Discussion

Isolated hypoglossal nerve paralysis is rarely encountered, particularly if there are no other associated signs and symptoms. Between 1990 and the beginning of 2000 only 20 papers relating to hypoglossal nerve palsy could be found. The majority of papers were published in neurological/neurosurgical or radiological journals. The few publications in ENT journals concerned with the 12th nerve paresis mainly described hypoglossal nerve schwannomas and other skull base tumours and their surgical management, respectively.

The 12th cranial nerve is mainly motor in function, although it is believed that a few proprioceptive fibres from the tongue pass through the hypoglossal nerve and terminate in the trigeminal nuclei.[1] If looking at the function of the nerve, it is important to distinguish between the 12th nerve proper and fibres of cervical origin which join it. These fibres originate from C1, first forming a sensory branch which supplies the posterior fossa dura; second, innervating the geniohyoid muscle; and third, giving off the superior root to the ansa cervicalis, also called ansa hypoglossi. Via the ansa cervicalis, the inferior root of which is formed by C2 and C3 fibres, all the infrahyoid muscles (sternohyoid, omohyoid, thyrohyoid and sternohyoid) are innervated. The 12th nerve proper exclusively supplies all intrinsic and extrinsic muscles of the tongue except palatoglossus which is supplied by the pharyngeal plexus.

Paralysis of the hypoglossal nerve results in compromise of the motor function of the tongue, with deviation of the tongue towards the side of the lesion if the lower motor neurone (nuclear and infranuclear disease) is affected, and away from the side of the lesion if the upper motor neurone (supranuclear disease) is involved. This can cause mild dysphagia and dysarthria, but most patients with an isolated paralysis compensate well. Bilateral paralysis results in more severe dysphagia and dysarthria. The severity of symptoms will also depend on the rapidity of onset and on the involvement of other lower cranial nerves.[1] However, it is surprising how minor the symptoms can be. In the largest reported series of hypoglossal nerve palsies, only 10 patients out of 100 (33 with a bilateral paresis) were admitted with complaints specifically related to tongue function. The author therefore considered 12th palsy as a sign rather than a symptom.[2] Patients may complain about fasciculation of the tongue, a phenomenon which only occurs if the lower neurone is affected. Approximately 4–6 weeks after the injury, the intrinsic and extrinsic muscles of the tongue undergo atrophy and fatty replacement, a process which eventually results in wasting of the tongue. Atrophy of the tongue should only be observed if the disorder is located at the nuclear or infranuclear level. Because of the close anatomical relationship to lower cranial and autonomic nerves, isolated 12th nerve neuropathies are unusual.

Because many diseases can affect the hypoglossal nerve along its path from the brain stem to the tongue, it is important to have a systematic approach in order to identify causes of palsy. This is achieved by looking at the nerve from topographic-segmental points of view as suggested by Thompson and Smoker.[3] The nerve is divided into five segments the medullary, cisternal, skull base, naso/oropharyngeal carotid space, and sublingual segments. Because each segment may be affected by different disorders, localizing a lesion to a particular segment allows a narrower differential diagnosis. The differential diagnosis is varied and the following disorders can affect the various segments of the nerve.

Medullary segment: infarct and haemorrhage (stroke), brain stem metastasis, brain stem glioma, demyelination (e.g. multiple sclerosis), motor neurone disease, infection (brain stem encephalitis), vascular malformations, syringobulbia, Chiari malformation.

Cisternal segment: chordoma and meningioma, nerve sheath tumours, meningeal metastases, meningitis (including tuberculosis and sarcoidosis), subarachnoid haemorrhage, disease of the vertebral arteries, rheumatoid arthritis (abnormal odontoid process).

Skull base segment: skull base metastases, nerve sheath and glomus tumours, nasopharyngeal carcinoma, plasmacytoma and multiple myeloma, primary bone tumours (sarcomas), dysplastic bone disease (Paget's, fibrous dysplasia), skull base osteomyelitis, trauma (skull base fracture).

Carotid space segment: oropharyngeal and nasopharyngeal carcinoma, salivary gland malignancies, lymphoma, lymph node metastases from head/neck tumours and distant sites, nerve sheath and glomus tumours, vascular disease (dissecting aneurysm, thrombosis), carotid space infection, radiation, operation (carotid endarterectomy, head and neck surgery), trauma (gun shot, stabbing).

Sublingual segment: carcinoma of the oropharynx and oral cavity, salivary gland malignancies, infection of the sublingual space, radiation.

Other conditions that may affect the 12th nerve but are difficult to allocate to a certain segment are viral infections such as infectious mononucleosis and herpes simplex, idiopathic multiple cranial neuropathies and Guillain–-Barre syndrome.

In the largest series of 100 hypoglossal nerve palsies,[2] nearly half of the cases were caused by tumours, and more than half of the tumours were malignant. It is not explicitly stated in this paper how many 12th nerve palsies occurred as an isolated phenomenon, but in the great majority other cranial nerves were involved. Metastasis, chordoma, nasopharyngeal carcinoma, and lymphoma composed nearly two-thirds of the tumours. Metastasis may involve the hypoglossal nerve within the medulla, in the subarachnoid space, at the skull base, within the hypoglossal canal, or in the neck. Occasionally, 12th nerve palsy is the sole presenting sign of a meningeal tumour. There are series cited in the same paper which demonstrate that hypoglossal nerve involvement is not uncommon due to meningeal metastases (11 per cent and 15 per cent involvement) and skull base metastases (nine out of 43 had a unilateral 12th nerve palsy). In this neurological case mix, nasopharyngeal tumours (nasophayngeal carcinoma, lymphoma, plasmacytoma) and metastatic carcinoma were the most common tumours. The conclusion was that 12th nerve palsy is an ominous sign. However, hypoglossal nerve palsy is uncommon in nasopharyngeal carcinoma at the time of the initial diagnosis: cranial nerve palsy is seen in 13–25 per cent of patients at diagnosis and is an adverse prognostic factor for patient survival.[4] The majority of cranial nerve palsies on presentation involve the abducens and trigeminal nerves. This is due to the propensity of nasopharyngeal carcinoma to spread superiorly into the sphenoid and through the skull base foramina, rather than in a postero-inferior direction to involve the basiocciput and the carotid sheath. A hypoglossal nerve palsy is a marker of advanced local disease seen only in a minority of patients.[4]

Radiation-induced cranial nerve palsies have been described following treatment for tumours of the neck, oral cavity, tongue and nasopharynx. In these cases, the 12th nerve is more commonly involved and the mechanism of damage is believed to be either direct, or indirect, secondary to vascular damage and fibrosis in the surrounding tissue.[5]

The pathogenesis in our case was pressure on, and stretching of, the nerve due to the intramural haematoma of the dissecting internal carotid artery (see Figures 1 and 2).

ENT surgeons may encounter a patient with an isolated 12th nerve palsy of unknown origin, if there are features on imaging which suggest a tongue lesion, as in our case. There are radiological features associated with denervation which can simulate a tumour within the tongue. The tongue muscles may undergo fatty change causing alteration in signal on MRI and density on CT.[3] Chong and Fan[6] state that at an early stage of hypoglossal nerve palsy the radiological signs of atrophy and fatty infiltration may be subtle, but the affected side of the tongue can be seen to drop backwards in axial images creating the impression of an apparent increase in its length. This could be mistaken for an increase in size of the tongue and they describe a case where a patient with a 12th nerve palsy underwent repeated biopsies of the tongue for a suspected tumour involvement.

Conclusion

The rare phenomenon of isolated hypoglossal nerve palsy can be the result of one of many pathologies that can affect the 12th nerve along its course from the brain stem to the tongue, in this case a non-traumatic dissecting aneurysm of the internal carotid artery. Careful review of imaging should be performed prior to biopsy of the tongue, as the denervated swollen tongue may mimic a tumour both clinically and radiologically.

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Address for correspondence:
Riad Shahab,
14 Galen Close,
Epsom KT19 7DL, UK.

E-mail: riadshahab@doctors.org.uk

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