LESSON OF THE MONTH

Transient total tongue paralysis from simultaneous central and peripheral lesions

M S Mouradian, K M Chan, T Jeerakathil, A Shuaib

Lower cranial nerve palsies in various combinations involving the hypoglossal, glossopharyngeal, vagal, and the accessory nerves occur in internal carotid artery (ICA) dissection.^{1–8 17} The hypoglossal nerve (cn-XII) is most often affected. It is suggested that the close proximity of these nerves to the cervical ICA may subject them to mechanical injury either by the expansion of the dissected artery or by aneurysm formation.^{1 3 9 10} Dissection of the ICA may cause transient or permanent disruption of the blood supply to these cranial nerves.^{6 11} Therefore, cranial nerve injuries may result from “mechanical, embolic, or haemodynamic” processes caused by ICA dissection.⁶ This hypothesis is supported by anatomical and clinical findings.^{2 6 12} Each half of the genioglossus muscle that protrudes the tongue is innervated by contralateral corticobulbar (crossed) fibres whereas all other intrinsic and extrinsic tongue muscles that move the tongue within the mouth in all directions have bilateral cortical innervation.^{13 14} Unilateral cortical lesion in the topography representing the tongue may therefore cause contralateral genioglossus muscle weakness, resulting in deviation of the protruded tongue away from the side of the cortical lesion. By contrast, cn-XII injury causes ipsilateral genioglossus muscle weakness; hence the protruded tongue deviates to the side of the injured nerve.

When clinical history is unrevealing and lower cranial nerves are involved, diagnosing ICA dissection becomes challenging. We present a patient with complete tongue paralysis that resulted from a frontal lobe stroke and a cn-XII injury, both caused by an ipsilateral ICA dissection. In the absence of binuclear cn-XII lesion or a pseudobulbar syndrome, localisation of the lesions responsible for the complete tongue paralysis can be difficult. This patient illustrates that in the presence of bilateral acute tongue paralysis with failure to protrude the tongue, the possibility of an ICA dissection with a simultaneous and ipsilateral cn-XII injury (peripheral lesion) and a frontal lobe stroke (central lesion) should be considered.

Case report

A 47 year old right handed white man with a history of heavy smoking was brought to the emergency room by his wife about 45 minutes

after the onset of left face, arm, and hand weakness, inability to speak, and difficulty with breathing. Except for a strong cough from a common cold, he had been in his usual state of health. At the emergency room the patient could not speak but could comprehend. He communicated by nodding his head and gesturing with his right hand. His right pupil was 2 mm in diameter and the left 3 mm, both reactive to light. Visual fields were full as tested by confrontation. The right eyelid was mildly ptotic. Left facial sensation to touch, pin prick, and temperature was decreased and there was a central type left facial palsy. Corneal reflex was decreased on the left side. He was unable to protrude his tongue beyond the front teeth, and he could not produce side to side tongue movements. Lowering the head of the stretcher to less than 30 degrees caused the tongue to flop back and entirely obstruct his upper airway. He was, however, able to produce sounds, shaping them with jaw and lip movements. Soft palate elevation was weaker on the right side. Gag reflex was present bilaterally, but he was unable to swallow his saliva. At the time of neurological evaluation (about 55 minutes after symptom onset) he could produce motor resistance with his left arm and hand against the examiner (motor strength was assessed as MRC grade 4/5). He had decreased sensation over his left arm and hand. The right side of his body and his left leg had normal motor and sensory functions. Stretch reflexes and cerebellar functions were normal.

His airway was promptly secured by a nasolaryngeal tube and a cerebral CT was obtained immediately. The scan disclosed early stroke changes in the right frontal lobe. Because of the Horner's sign on the right side, a carotid angiogram was performed (at 2 hours into his stroke). The angiogram disclosed dissection of the right ICA with occlusion of the vessel (fig 1). The collateral supply to the right hemisphere was primarily through the right posterior communicating artery and collateral supply to the right anterior cerebral artery territory was through the anterior communicating artery. The patient was then started on heparin which was subsequently changed to warfarin for the next 3 months.

By the second day the left facial palsy started to improve and his tongue stopped falling back. He no longer needed a nasopharyngeal tube to

Division of Neurology,
University of Alberta
Hospital, 2E3 13
Mackenzie Health
Sciences Centre, 8440
112 Street, Edmonton,
Alberta T6G 2B7,
Canada
M S Mouradian
K M Chan
T Jeerakathil
A Shuaib

Correspondence to:
Professor A Shuaib
ashfaq.shuaib@ualberta.ca

Received 14 August 2000
and in revised form
17 January 2001
Accepted 18 January 2001

secure the airway. He could protrude his tongue, which deviated to the left. His speech was severely dysarthric. A repeat CT obtained on day 2 showed an area of decreased cortical and subcortical attenuation in the right frontal lobe, compatible with a right frontal lobe stroke (fig 2). On day 7, his tongue still deviated to the left when protruded, but the side to side tongue movements inside the mouth were stronger than before. His dysarthria was now moderate and he was able to drink thickened liquids. On day 8 the patient was transferred to a rehabilitation unit. On day 45, at follow up assessment his tongue continued to deviate to the left upon protrusion. His left face and arm had a moderate sensory loss and weakness. His tongue did not show fasciculations.

Figure 1 Angiogram showing right ICA dissection with beak-like tapering and occlusion.

Figure 2 Brain CT showing a small right frontal infarct.

An EMG examination of the tongue muscles was performed 2 weeks after the acute event. Needle EMG examination showed impaired recruitment in the left orbicularis oculi and masseter muscles. With the concentric needle electrode inserted into the genioglossus muscle through the base of the chin, no fibrillation potentials, positive sharp waves, or fasciculation potentials were seen on either side and insertional activities were normal. However, voluntary recruitment in the left genioglossus muscle was impaired.

Discussion

To our knowledge this is the first reported case of bilateral transient tongue paralysis caused by two tandem lesions, one central and the other peripheral. Involvement of lower cranial nerves in ICA dissection may cause false clinical localisation of the lesion.¹⁵ The above presentation of tandem lesions may further complicate the clinical localisation and delay definitive diagnostic procedures and the treatment options. Especially, when the oculosympathetic signs of ICA dissection are not obvious, the presence of cranial nerve dysfunction may deceptively suggest brain stem pathology.

Unilateral cortical lesions produce contralateral tongue weakness causing the protruded tongue to deviate away from the side of the central lesion. Contrary to this, unilateral cn-XII lesion causes the protruded tongue to deviate to the side of the lesion. To cause simultaneous bilateral tongue weakness with a single peripheral nerve lesion, both nuclei of the cn-XII need to be affected at the same time. This is possible, as, in the tegmentum of the upper medulla oblongata, both of these nuclei are in close proximity to each other.¹⁴ Multiple sclerosis or syringobulbia, and possibly other destructive lesions may affect both cn-XII nuclei.¹⁶ Bilateral paralysis of the genioglossus muscle may develop (1) in the presence of two lesions simultaneously affecting the corticobulbar fibres bilaterally, (2) in the presence of bilateral cn-XII lesion, or (3) two ipsilateral tandem lesions. Our patient experienced the last possibility.

His right sided ICA dissection most likely injured the right cn-XII (peripheral lesion), by the possible mechanisms mentioned in the introduction, giving rise to severe right sided genioglossus muscle weakness. The carotid dissection also caused a right hemispheric thromboembolic stroke affecting the sensorimotor areas representing the left face, the tongue, left half of genioglossus muscle, and the left arm and hand. The right cortical stroke (the central lesion) disrupted the motor function of the left genioglossus muscle most severely owing to this muscle's crossed unilateral (contralateral) cortical innervation. The simultaneous occurrence of two ipsilateral lesions—one peripheral, affecting the right cn-XII, and the other, central affecting the motor cortex on the right controlling the left genioglossus muscle—culminated in a total paralysis of tongue protrusion. The recovery of the motor strength of the right side of the tongue was clinically evident within days. The lack of any sign of denervation on needle EMG examination performed 2 weeks after the ICA

dissection, along with the rapid functional recovery, suggest that the weakness on the right side of the tongue was probably due to a neurapraxic injury resulting in a conduction block, which recovered quickly. The reduced recruitment on the left side of his tongue most likely reflected impaired central motor input as a result of the right cortical stroke. Although a neurapraxic injury to the left cn-XII nucleus/nerve fibres could have been an additional cause of tongue weakness, mechanistically it is much less likely.

- 1 Lane RJ, Weisman RA, Savino PJ, et al. Aneurysm of the internal carotid artery at the base of the skull; an unusual cause of cranial neuropathies. *Otolaryngol Head Neck Surg* 1980;**88**:230–2.
- 2 Havelius U, Hindfelt B, Brismar J, et al. Carotid fibromuscular dysplasia and paresis of lower cranial nerves (Collet-Sicard syndrome). Case report. *J Neurosurg* 1982;**56**:850–3.
- 3 Goodman JM, Zink WL, Cooper DF. Hemilingual paralysis caused by spontaneous carotid artery dissection. *Arch Neurol* 1983;**40**:653–4.
- 4 Panisset M, Eidelman BH. Multiple cranial neuropathy as a feature of internal carotid artery dissection. *Stroke* 1990;**21**:141–7.
- 5 Mokri B, Schievink WI, Olsen KD, et al. Spontaneous dissection of the cervical internal carotid artery. Presentation with lower cranial nerve palsies. *Arch Otolaryngol Head Neck Surg* 1992;**118**:431–5.
- 6 Mokri B, Silbert PL, Schievink WI, et al. Cranial nerve palsy in spontaneous dissection of the extracranial internal carotid artery. *Neurology* 1996;**46**:356–9.
- 7 Leys D, Lucas C, Gobert M, et al. Cervical artery dissections. *European Neurol* 1997;**37**:3–12.
- 8 Boukobza M, Ast G, Reizine D, et al. Internal carotid artery dissection causes hypoglossal nerve palsy: CT, MRI, and angiographic findings. *J Neuroimaging* 1998;**8**:244–6.
- 9 Scotti G, Melancon D, Oliver A. Hypoglossal paralysis due to compression by tortuous internal carotid artery in the neck. *Neuroradiology* 1978;**14**:263–5.
- 10 Fisher CM, Ojemann RG, Roberson GH. Spontaneous dissection of cervico-cerebral arteries. *Can J Neurol Sci* 1978;**5**:9–19.
- 11 Gobert M, Mounier-Vehier F, Lucas C, et al. Cranial nerve palsies due to internal carotid artery dissection: seven cases. *Acta Neurologica Belgica* 1996;**96**:55–61.
- 12 Lapresle J, Lasjaunias P. Cranial nerve ischemic arterial syndromes. A review. *Brain* 1986;**109**:207–16.
- 13 Riggs JE. Distinguishing between extrinsic and intrinsic tongue muscle weakness in unilateral hypoglossal palsy. *Neurology* 1984;**34**:1367–8.
- 14 Haerer AF. The hypoglossal nerve. In: *DeJong's The Neurologic Examination*, 5th ed. Philadelphia; JB Lippincott, 1992: 251–7.
- 15 Hess DC, Sethi KD, Nicholas FT. Carotid dissection: a new false localising sign [letter]. *J Neurol Neurosurg Psychiatry* 1990;**53**:804–5.
- 16 Brazis PW, Masdeu JC, Biller J. The localization of lesions affecting cranial nerve xii (the hypoglossal nerve). In: Brazis PW, Masdeu JC, Biller J, eds. *Localization in clinical neurology*, 2nd ed. Boston; Little, Brown, 1990:259–67.
- 17 Maitland CG, Black JL, Smith WA. Abducens nerve palsy due to spontaneous dissection of the internal carotid artery. *Arch Neurol* 1983;**40**:448–9.