CASE REPORT

A rare presentation of spontaneous internal carotid artery dissection with Horner’s syndrome, VIIth, Xth and XIth nerve palsies

Azer Majeed1,*, Nuno Pedro Lobato Ribeiro1,†, Asem Ali2,†, Mohsen Hijazi1,† and Hina Farook3,†

1Stroke Unit, Scunthorpe General Hospital, Scunthorpe, UK, 2Stroke Unit, Diana Princess of Wales Hospital, Grimsby, UK, and 3Medicine, Rawalpindi Medical College, Rawalpindi, Pakistan

*Correspondence address. Stroke Unit, Scunthorpe General Hospital, Cliff gardens Scunthorpe DN157BH, UK. Tel: +00447572792913; E-mail: azermajeed@gmail.com

Abstract

Spontaneous internal carotid artery dissection (sICAD) is an uncommon cause of isolated cranial nerve palsies. Commonly patients present with stroke, headache, facial pain and Horner’s syndrome, with upto 16% having cranial nerve palsies. We present the case of a 55-year-old man who presented with hoarseness, dysphagia and tongue swelling, mimicking a tongue base tumor. He was found to have unilateral VIIth, Xth and XIIth nerve palsies with Horner’s syndrome. Magnetic resonance imaging showed high signal changes and loss of signal void in right internal carotid artery, later confirmed by Angiography as a dissection with pseudo-aneurysm. He was started on anticoagulation and made a good recovery on discharge. This case presents a unique combination of cranial nerve palsies due to internal carotid artery dissection (ICAD) and to our knowledge is the first reported case in the literature. Early recognition and institution of appropriate therapy is critical to prevention of ischemic stroke.

INTRODUCTION

Spontaneous internal carotid artery dissection (sICAD) is known to cause upto 2% of all ischemic strokes and occurs most frequently in the fifth decade of life [1]. It can be due to local trauma or be spontaneous (sICAD). It presents most commonly as stroke (67%) while Horner’s syndrome is seen in 28–58% of cases [1]. Cranial nerves are involved in 8–16% of cases, with lower cranial nerves IX–XII most commonly affected [1]. We present, to our knowledge, the first reported case in the literature of Horner’s syndrome with VIIth, Xth and XIIth cranial nerve palsies due to sICAD.

CASE PRESENTATION

A 55-year-old man, with past history of hypertension, self-presented to the emergency department 3 days after acute onset of frontal headache followed by hoarseness of voice and dysphagia. His blood pressure (BP) was 189/138 mmHg and a plain computed tomography (CT) head was done which was unremarkable. He was admitted under the ENT department and a Naso-endoscopy was done the following day which showed right vocal cord paralysis (VCP) (Xth nerve) with para median pooling (Fig. 1), swelling and paralysis of
right half of tongue (XIIth) decreased movement of right half of pharyngeal wall but with intact pharyngeal sensations. A provisional diagnosis of tumour of the base of the tongue was made and CT Neck–Thorax–Liver with contrast was done. The CT scan only showed thickening of right half of the tongue base and right lateral pharynx, with no other pathology. Subsequently, MRI head was requested and the Stroke team was called to review the patient. On detailed neurological examination, in addition to the tongue base swelling and deviation towards the right, he also had right partial ptosis, right miosis (1 mm) and right lower motor neuron type Facial palsy (VIIth). He had nasolabial fold flattening and decreased wrinkling of forehead on the right half of face however lacrimation, taste and hearing were unaffected. There was no limb motor or sensory deficit present. MRI head and neck with contrast showed T1 and T2 intramural high signal changes with loss of normal signal void in the right ICA (arrow). A diagnosis of spontaneous right internal carotid artery dissection (ICAD) was made as there was no history of antecedent trauma. Anticoagulation was started with warfarin and a CT angiogram was requested for a detailed look at the carotid arteries. The CT angiogram confirmed dissection of right ICA just before its intraosseous part with pseudo-aneurysm formation (Fig. 4).

The patient was discharged home with a therapeutic INR (between 2 and 3), with a view to continue warfarin for 3–6 months. He received speech and language therapy during the admission and his BP was controlled with Amlodipine, Ramipril and Bendroflumethiazide. A follow-up CT angiogram has been arranged in 12 weeks’ time with neurovascular follow-up. On 6 weeks review visit, his speech and swallowing had recovered to normal although the partial Horner’s syndrome and facial palsy had not completely improved (Figs 5 and 6).

**DISCUSSION**

Spontaneous ICAD has been reported to cause various combinations of cranial nerve palsies involving upper as well as lower cranial nerves. Three syndromes have been named in the literature, describing the involvement of multiple lower cranial nerves (IX–XII) in different combinations with ICAD as a possible cause (Table 1) [2].
Two of these syndromes, Villaret’s and Collet Sicard, have been previously reported in the literature as a consequence of sICAD [3, 4]. Tapia’s syndrome, first described by a Spanish Otolaryngologist Antonio Garcia Tapia in 1904, has been variously interpreted by different authors and there is little agreement currently on the symptoms included in this syndrome.

The most frequently involved nerves are XIth, Xth and Xth [5]. VIIth nerve palsy due to sICAD is rare but cases of isolated Facial palsy have been reported in the literature [6]. Panisset et al. reported a case of left ICAD after head injury in 1990 with Vth, VIIth, IXth, Xth and XIIth nerve abnormalities [7], and although Facial, vagus and hypoglossal nerves were involved in that case too but there were no features of Horner’s syndrome, as was present in our case.

The lower cranial nerves IXth, Xth and XIIth leave the skull through the jugular foramen, while the XIIth nerve exits through the hypoglossal canal just inferiorly. In their early course, all four of these cranial nerves lie close together near the carotid artery, peri-carotid sympathetic plexus and jugular vein with cranial nerves X and XII having the longest anatomic relationship to this artery [8]. In most cases, the dissection begins distal to carotid bulb and shows maximal distension at C1 or C2 vertebral level [5]. At this point, the IXth cranial nerve lies lateral to ICA while the Xth lies posteriorly. Hypoglossal crosses the ICA more caudally at the C3 level [5] [Fig. 7]. Facial (VIIth) nerve palsy can be seen if the dissection extends into the retroparotid space [9]. Compression of the middle meningeal artery by the expanding dissected ICA can also lead to VIIth nerve ischemia, and in some cases disruption of an anatomically variant facial supply from ICA may be the cause of Facial palsy as suggested by Fioravanti et al. [6].

Another rare feature of sICAD is VCP, which was a prominent feature of our case. Although the most common causes of VCP are extra laryngeal tumours, surgical insult and trauma, but almost 16% of cases with sICAD also have VCP [10]. Similarly, unilateral tongue swelling, as seen in our case, can be an early presentation of sICAD with VIIth nerve palsy and can mimic angioedema. Transient unilateral denervation causing fluid shift into extracellular space can lead to oedema and tongue swelling. ICAD has encouraging prognosis and in 70–80% of cases complete or near complete recovery is seen [1]. Currently both Antiplatelet and Anticoagulation are considered equally effective conservative modes of treatment. Endovascular and surgical methods are reserved for those who fail to respond to medical therapy [1].

sICAD can present as different combinations of cranial nerve palsies and should be considered as a differential diagnosis of VIIth, Xth and XIIth nerve palsies with Horner’s syndrome. Tongue swelling and VCP due to it can mimic tumours but a detailed neurological examination and early Angiography can help in prompt diagnosis.

**Table 1:** Syndromes with multiple lower cranial nerves palsy due to ICAD

| Syndrome               | Lesion location               | Structures involved                      | Clinical findings                                      | Comment                                                                 |
|------------------------|------------------------------|-----------------------------------------|--------------------------------------------------------|------------------------------------------------------------------------|
| Collet Sicard (MacKenzie, Lannoois-Jouty) | Posterior lateral condylar space | CN IX;XI;XII; Carotid sympathetics      | Weakness in the distribution of involved nerves.       | Neoplasm of skull base, glomus jugulare, carotid aneurysm (including dissection) |
| Villaret’s              | Retropharyngeal space         | CN IX;XII; Carotid sympathetics         | Weakness in the distribution of involved nerves.       | Neoplasm of skull base, glomus jugulare, carotid aneurysm (including dissection) |
| Tapia’s                 | Retroparotid space            | CN X;XI;XI; Carotid sympathetics        | Weakness in the distribution of involved nerves; Horner’s syndrome | Usually due to tumour of parotid or skull base, occasionally carotid aneurysm (including dissection) |

**Figure 4:** CT Angiogram above arch of aorta showing dissection of right ICA just before its intraosseous part with a pseudo-aneurysm (arrow).

**Figure 5:** Follow-up visit showing partial resolution of Horner’s syndrome and improved forehead wrinkling.

**Figure 6:** Follow-up visit showing partial improved right nasolabial fold flattening.
ACKNOWLEDGEMENTS
None.

CONFLICT OF INTEREST STATEMENT
None declared.

FUNDING
This work has been carried out without any grants or funds.

ETHICAL APPROVAL
Not needed.

CONSENT
Informed consent has been taken from the patient himself.

GUARANTOR
Dr Azer Majeed.

REFERENCES
1. Fusco MR, Harrigan MR. Cerebrovascular dissections—a review Part I: spontaneous dissections. Neurosurgery 2011;68:242–57.
2. Campbell WW. Dejong’s the Neurologic Examination. Philadelphia: Lippincott Williams & Wilkins, 2005, 291–2.
3. Battaglia F, Martini L, Tannier C. Collet-Sicard syndrome after carotid artery dissection. Rev Neurol (Paris) 2009;165:588–90.
4. Mizutani S, Tsukuura R, Matsumura K, Watanabe M, Hanakawa I, Kamata T. Villaret’s syndrome caused by internal carotid artery dissection. Rinsho Shinkeigaku 2011; 51:608–11.
5. Sturzenegger M, Huber P. Cranial nerve palsies in spontaneous carotid artery dissection. J Neurol Neurosurg Psychiatry 1993;56:1191–9.
6. Fioravanti V, Vinceti G, Chiari A, Canali E, Frigio Nichelli P, Mandrioli J. Internal carotid artery dissection: a rare cause of peripheral facial nerve palsy. Eur Neurol 2012; 68:74.
7. Panisset M, Eidelman BH. Multiple cranial neuropathy as a feature of internal carotid artery dissection. Stroke 1990;21:141–7.
8. Mokri B, Silbert PL, Schievink WI, Piepgras DG. Cranial nerve palsy in spontaneous dissection of the extracranial internal carotid artery. Neurology 1996;46:356–9.
9. Bone J, Hadley D. Syndromes of the orbital fissure, cavernous sinus, cerebellar pontine angle, and skull base. J Neurol Neurosurg Psychiatry 2005;76(Suppl 3):iii29–38.
10. Nguyen TJ, Zhang H, Dziegielewski PT, Seemann R. Vocal cord paralysis secondary to spontaneous internal carotid dissection: case report and systematic review of the literature. J Otolaryngol Head Neck Surg 2013;42:34.