CASE REPORT

Collet-Sicard syndrome as a result of unilateral carotid artery dissection

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SUMMARY

A 52-year-old man presented with sudden onset symptoms of multiple cranial nerve palsies (IX, X and XII) following a 10-day history of coryzal illness. Follow-up examination established atrophy of the trapezius suggesting additional involvement of the spinal accessory nerve (XI). Further investigation including CT and MRI demonstrated dissection of the internal carotid artery. Given the involvement of cranial nerves IX to XII, and that the patient demonstrated no signs of Horner's syndrome, we suggested that this patient fits the description of Collet-Sicard syndrome. On vascular opinion the carotid dissection was expected to resolve without intervention but the patient was prescribed aspirin for thromboprophylaxis. Gradual resolution of neurological symptoms was observed at 8-week follow-up.

BACKGROUND

Cranial nerve palsies are a common symptom of internal carotid artery dissection (ICAD).¹ In contrast to many previously reported cases that demonstrate isolated single cranial nerve palsies² or combinations of several lower cranial nerve palsies³ as a result of ICAD, our case illustrates a broad range of symptoms due to IX, X, XI and XII cranial nerve involvement without any features of cerebral ischaemic change. The complete set of lower cranial nerve palsies from IX to XII and sparing of the sympathetic plexus demonstrated by our patient fits the description of a rare clinical syndrome most commonly caused by trauma and skull base neoplasms.⁴ Hence, we present this unusual case of Collet-Sicard syndrome as a result of ICAD, most likely as a result of the minor trauma associated with coughing during coryzal illness.

CASE PRESENTATION

A 52-year-old man was referred to the ear, nose and throat department from general practice, with a history of sudden onset left facial pain, ipsilateral tongue weakness, dysphonia, dysphagia, regurgitation of solid food and altered sensation of taste. The patient described waking up with the symptoms following a 10-day history of coryzal illness. The patient has an umbilical hernia but no other medical history of note, is a non-smoker and drinks approximately 16–20 units of alcohol per week.

On initial examination, the patient was found to have atrophy of the left glossal musculature and an accompanying left hypoglossal nerve paresis, in addition to a left vocal cord palsy. The patient demonstrated no signs of Horner's syndrome and examination of the neck showed no abnormalities.

At follow-up 1 month later a degree of atrophy of the left trapezius was apparent and the glossopharyngeal nerve was found to be intact with bilateral gag reflex.

INVESTIGATIONS

Initial axial CT scan from head to thoracic inlet was inconclusive but further investigation including MRI (see figure 1) demonstrated dissection of the internal carotid artery. Brain MRI did not demonstrate any ischaemic change. Laryngoscopy showed a left vocal cord palsy.

TREATMENT

Subsequent to investigations, the patient was referred to a consultant in general medicine who sought additional advice from the vascular team on how to manage the dissection. No active vascular treatment was considered necessary with the carotid dissection expected to resolve without intervention. The patient was initially started on 300 mg aspirin reducing to a dose of 75 mg daily for a minimum of 6 months for thromboprophylaxis.

Figure 1 Axial fat saturation T1-weighted MRI (taken 5 weeks after onset of symptoms) demonstrating a crescent-shaped area of intramural haematoma (arrow) as a consequence of left carotid artery dissection. Although not obvious to the eye, measurement of the left carotid lumen showed narrowing compared with the right side, a further characteristic of dissection.

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OUTCOME AND FOLLOW-UP

The patient was referred to the neurology department for further follow-up. Gradual improvement of neurological symptoms was observed at 8-week follow-up. The patient continues to be seen by the neurology team to monitor further improvement of symptoms.

DISCUSSION

Collet-Sicard syndrome (CSS) is a rarely seen condition characterised by palsies of the lower cranial nerves IX to XII with sparing of the sympathetic plexus.^{5–6} There are several known causes of CSS, the most common of which are trauma and primary neoplasms of the ear and skull base.⁴ Lesser seen causes include metastases, jugular phlebitis, polyarteritis nodosa, Lyme disease and, as in the case of our patient, ICAD.⁷

Carotid artery dissection is most often seen as a result of trauma with injury commonly sustained through the rapid deceleration seen in road traffic accidents.⁷ Dissections without a major traumatic cause are poorly understood, although connective tissue disorders such as Ehlers-Danlos syndrome and Marfans disease have been implicated.⁸ It has also been suggested that upper respiratory tract infection could be a catalyst for cervical artery dissections as a result of the minor trauma of coughing and sneezing, causing direct mechanical stress to the artery.⁹

Only a handful of cases describing carotid artery dissection without major traumatic injury as a cause of CSS have been published.^{10–11} We believe our case to be unique in that we identify the minor trauma associated with upper respiratory tract infection as the likely cause of the dissection in this patient given his 10-day history of common cold symptoms preceding the onset of neurological symptoms and the absence of major traumatic injury.

Two possible mechanisms of cranial nerve involvement in carotid dissection have been described.¹² The mechanical compromise resulting from dissection can impede blood flow through the nutrient vessels supplying the cranial nerves. Although anatomical variations are possible, the site of the dissection determines which nutrient vessels are haemodynamically compromised. Given that the external carotid artery supplies the lower cranial nerves through the ascending pharyngeal system we deem this an unlikely mechanism as the dissection in our patient occurred in the internal rather than the external carotid artery.

Alternatively, intramural haematoma can cause the artery to expand and directly compress the cranial nerves (IX, X, XI, XII) running through the upper carotid sheath. Although we cannot definitively rule out ischaemic damage to the cranial nerves, this is the more likely mechanism given the dissection of the internal carotid and the accompanying lower cranial nerve palsies that our patient demonstrates:

- ▶ Glossopharyngeal (IX): reported difficulty swallowing and loss of taste sensation.
- ▶ Vagus (X): left vocal cord palsy observed on flexible laryngoscopy.
- ▶ Spinal accessory (XI): atrophy of trapezius established on examination at monthly follow-up.
- ▶ Hypoglossal (XII): ipsilateral tongue paresis.

The common site of convergence of all the lower cranial nerves is the retrostyloid section of the parapharyngeal space. The posterior boundary of this space is the fascia of the carotid sheath, hence explaining how an intramural haematoma of the carotid artery can directly compress the lower cranial nerves to

manifest clinically as multiple palsies. Interestingly, our patient does not demonstrate any signs of Horner's syndrome which could also be expected as sympathetic fibres also pass through the parapharyngeal space. Such sparing of the sympathetic plexus provides further argument for the classification of this case as CSS.

However, we do exercise a degree of caution in definitively describing this as a case of CSS given that the patient was found to have an intact bilateral gag reflex. The vagus nerve palsy could also account for the patients reported difficulty in swallowing. Despite this we feel that the combination of difficulty in swallowing and the altered sense of taste are suggestive enough of glossopharyngeal nerve palsy for the classification of CSS to be given serious consideration.

This case should serve as an important reminder that carotid artery dissection can present with a broad range of symptoms as a result of multiple cranial nerve palsies. This case also highlights that carotid dissection should be suspected in patients with neurological symptoms and with a history of upper respiratory tract infection due to the mechanical trauma of coughing and sneezing.

Learning points

- ▶ Carotid artery dissection should be considered as a differential diagnosis in patients presenting with multiple cranial nerve palsies.
- ▶ Thorough clerking including history of upper respiratory tract infection and trauma may identify risk factors for carotid dissection.
- ▶ Successful diagnosis and management of patients with cranial nerve palsies as a result of carotid dissection requires input from a variety of specialties.
- ▶ Suspicion of carotid artery dissection causing cranial nerve palsy warrants MRI to identify the dissection.

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Competing interests None.

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