CASE REPORT

Case of idiopathic isolated unilateral hypoglossal nerve palsy with spontaneous recovery

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SUMMARY

Isolated hypoglossal nerve palsy is uncommon because of its intimate relationship with the other lower cranial nerves. Keane reported that tumours, predominantly malignant, were the most common cause of hypoglossal nerve palsy. We report a case of isolated idiopathic unilateral hypoglossal nerve palsy in a 45-year-old Caucasian male where no cause could be identified despite extensive investigations. There was uncertainty around prognosis at onset due to the rarity of this condition. In the absence of a cause, an early referral to the speech and language therapist was made and interestingly our patient made an almost complete recovery within 18 months of onset. In a small case series, it has been reported that though rare, idiopathic hypoglossal nerve palsy has an excellent outcome in most cases, similar to the more common idiopathic seventh cranial nerve palsy (Bell's Palsy). We recommend an early referral for physiotherapy in such cases.

BACKGROUND

Isolated unilateral hypoglossal nerve palsy (HNP) is rare and is often considered a harbinger of malignancy and poses a diagnostic challenge.¹⁻⁴ The purpose of this case report is to raise awareness of this condition and discuss the common causes and investigations for this rare condition. This case is also a reminder that idiopathic HNP (where no cause can be identified) has a favourable outcome and this information could be used to reassure patients who develop this rare condition. In this case, our patient with idiopathic HNP made an almost complete recovery.

CASE PRESENTATION

A 45-year-old Caucasian male presented with a 2-week history of pain in the left side of the neck and noted his tongue pulled to the left side making eating, drinking and speaking difficult. He is normally fit and active, never smoked and has no significant medical history of note. He does not take any regular medications and there was no history of recent neck injury. On examination, the only neurological abnormality noted was the deviation of the tongue to the left with associated wasting of the left half of the tongue ([figure 1](#)), findings consistent with a unilateral lower motor neuron hypoglossal nerve (CN XII) palsy. Other cranial nerves were intact with no Horner's or pyramidal signs, no neck stiffness and a negative Kernig's sign. He also had a normal cardiovascular system examination.

INVESTIGATIONS

Initial investigations comprising an ECG, chest X-ray, full blood count, electrolytes, renal function, blood sugar, inflammatory markers and liver function tests were normal. In view of the left-sided neck pain, an urgent CT carotid angiogram was performed to look for internal carotid dissection/aneurysm, which was normal. Further investigations including connective tissue screen, lupus and vasculitic screen, HIV serology, syphilis serology and vitamin B12 were all normal. He underwent a formal ear, nose and throat (ENT) specialist assessment including a flexible pan-nasendoscopy which was unremarkable. A MRI of the head and neck with a magnetic resonance angiography (MRA) was arranged for further evaluation. These showed no abnormality, apart from two cystic lesions in the left lobe of the thyroid, which were sampled and showed no malignancy.

DIFFERENTIAL DIAGNOSIS

Isolated HNP can represent a diagnostic challenge.⁵ Our patient had no history of recent neck injury, head and neck surgery or radiotherapy, all of which have a predilection for hypoglossal nerve damage, either directly or indirectly via vascular ischaemia or fibrosis in the surrounding tissue.³ Our patient was non-diabetic, and immunological and serological tests did not show any evidence of connective tissue disease, infection or vasculitis. He had no signs of meningeal irritation and a formal ENT assessment was unremarkable. Imaging of the head and neck, including the vascular system, showed no abnormality to account for HNP. A diagnosis of idiopathic unilateral HNP was made as no cause could be identified.

TREATMENT

In the absence of a cause to account for the HNP, no definitive treatment could be offered. The neurologist was unable to give a prognosis and warned the patient of the possibility of the neurological deficit remaining permanent. As the patient was finding the tasks of speaking, eating and drinking increasingly difficult, an early referral to the speech and language therapist (SALT) was made who recommended specific regular exercises.

OUTCOME AND FOLLOW-UP

Seven months after the initial presentation, the patient reported a significant subjective improvement of his symptoms. When he was last reviewed in clinic, 16 months after the initial presentation, the patient had made an almost full recovery.

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Figure 1 Left-sided lower motor neuron hypoglossal nerve palsy.

DISCUSSION

Isolated unilateral HNP is rare and poses a diagnostic challenge. When obtaining a focused patient history, it is important to enquire about recent head and neck surgery, neck trauma, underlying malignancy and radiotherapy, all of which have a predilection for hypoglossal nerve injury. The most common iatrogenic causes reported for hypoglossal nerve injury are carotid endarterectomy and submandibular gland excision.³ The presence of neck pain should raise the suspicion of internal carotid artery dissection, so a CT-carotid angiogram, and MRI and MRA of the head and neck are useful initial investigations. CT and MR studies could also help in identifying any vascular abnormality, syringobulbia, Arnold-Chiari malformation, bony lesions or tumours of the base of skull. MRI allows direct visualisation of the different segments of the hypoglossal nerve and generally is the investigation of choice for evaluation of a patient presenting with HNP of unknown cause.⁶ However, MR studies are inherently time consuming, whereas CT imaging has a practical advantage of greater availability and could swiftly identify potentially serious or life-threatening conditions such as an acute vascular event or mass effect from a space occupying lesion in patients presenting with unexplained cranial nerve palsies. We therefore recommend CT imaging as the first investigation for evaluation of patients presenting with HNP. A negative CT study should be followed by an MRI and MRA in the diagnostic workup, which allows assessment of the entire course of the hypoglossal nerve.³

Hypoglossal nerve could also be affected in multiple sclerosis from demyelination involving the medulla. Similarly, rheumatoid arthritis and polyangiitis with granulomatosis (Wegener's granulomatosis) are rare causes for HNP. In a case series of isolated HNPs reported by Sharma *et al*,⁷ tubercular meningitis

was responsible for a third of the cases, all of whom had neck stiffness and Kernig's sign at presentation along with a history of low-grade fever. Their MRI head showed leptomeningeal enhancement and exudates suggesting meningeal involvement. Another study⁸ has reported transient HNP after oropharyngeal intubation for general anaesthesia, especially after orthopaedic and otolaryngology operations. HNP in these cases was due to hypoglossal nerve compression and overstretching, a phenomenon more commonly seen in males given they have larger hyoid bone dimensions compared with females.⁹ In this case series, the recovery rate in corticosteroid treated patients was similar to non-treated patients. This contrasted with an earlier study, where a short course of corticosteroid was reported to accelerate recovery.⁷ Our patient, however, made a full recovery without any corticosteroid therapy.

In the absence of trauma, recent surgery and radiotherapy, about 50% of the cases of isolated HNP are idiopathic and, like Bell's Palsy of the seventh cranial nerve, is transient and reversible with an excellent outcome in most cases.^{3,4}

Our case is a reminder for clinicians that, though rare, idiopathic isolated HNP has a favourable prognosis and an early referral to the SALT should be considered.

Learning points

- ▶ Isolated hypoglossal nerve injury is rare and warrants a detailed history and clinical assessment.
- ▶ MRI with magnetic resonance angiography of head and neck are useful initial investigations as it allows assessment of the entire course of the hypoglossal nerve.
- ▶ Idiopathic hypoglossal nerve palsy, where no cause can be identified carries a good prognosis.
- ▶ Early referral to the speech and language therapist should be considered in such cases.

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