An uncommon cause of isolated hypoglossal nerve palsy: a case report

Précis

A rare case of isolated hypoglossal nerve palsy observed clinically as tongue deviation, and diagnosed by MRI identification of a meningioma in the hypoglossal canal.

Abstract

Introduction: Isolated hypoglossal nerve (CNXII) palsy is rare due to the course of the nerve and its close proximity to other cranial nerves and vessels. Aetiology includes space-occupying lesions, head and neck trauma, and infections. Characteristically, hypoglossal nerve palsy presents with unilateral atrophy of tongue musculature and deviation of the tongue to the affected side.

Case report: A 50-year-old woman attended Belfast Dental Hospital complaining of tenderness and swelling of the left side of her tongue for the previous eight weeks. Her medical history was unremarkable and she was a non-smoker. On examination, there was no evidence of lymphadenopathy, asymmetry or swelling. Cranial nerves (CN) I-XI were intact; however, testing of CNXII revealed fasciculation and deviation of the tongue to the left on protrusion. A magnetic resonance imagery (MRI) scan revealed a lesion in the left hypoglossal canal, in keeping with a meningioma. The patient has now been referred to neurology, awaiting the possibility of neurosurgery.

Discussion: Hypoglossal nerve palsy is uncommon and rarely presents in isolation. It raises suspicion of a sinister underlying pathology and therefore a prompt referral for an MRI scan was made. Meningiomas arising in the hypoglossal canal are extremely rare and this is the fourth case to be reported in the literature.

Conclusion: Isolated hypoglossal nerve palsy can present in any clinical situation, either as a complaint or incidental finding. It highlights the importance of detecting subtle intraoral clinical changes, the use of appropriate imaging, and the importance of multidisciplinary teamwork in diagnosis and management of complex cases.

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Introduction

The hypoglossal nerve (twelfth cranial nerve; CNXII) is a motor nerve that is responsible for the innervation of all the extrinsic and intrinsic muscles of the tongue with the exception of the palatoglossus muscle. It originates from the hypoglossus nucleus in the medulla oblongata, passes laterally across the posterior cranial fossa, within the subarachnoid space, and exits the cranium via the hypoglossal canal. The nerve passes downwards and forwards in the neck between the internal carotid artery and the internal jugular vein until it reaches the lower border of the posterior belly of digastric muscle. Here it turns forward and crosses the internal and external carotid arteries and the loop of the lingual artery. In the floor of the mouth, it is intimately associated with the hyoglossus, the lingual nerve and duct of the submandibular gland, before continuing to move in an anterior direction to enter the tongue.[1]

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FIGURE 1a and 1b: Patient's tongue showing deviation to the left side on protrusion.

Table 1: Differential diagnoses for hypoglossal nerve palsy

Nasopharyngeal carcinoma
Metastatic disease to base of skull
Periostitis of hypoglossal canal
Head and neck trauma
Epstein-Barr virus infection
Dural arteriovenous fistula of the transverse sinus
Post-vaccination cranial neuritis
Carotid artery dissection/aneurysm
Post-retropharyngeal infection
Schwannoma
Meningioma
Idiopathic

Hypoglossal nerve palsy is uncommon due to its complex course and close proximity to other cranial nerves and vessels. Numerous causes of hypoglossal nerve palsies have been documented in the literature (**Table 1**). These include: nasopharyngeal carcinomas;[2] metastasis to the base of the skull;[3] trauma;[4,5] vertebral artery and extracranial internal artery dissection;[6-8] hypoglossal schwannoma;[9] Epstein-Barr virus infection;[10] and, post-vaccination cranial neuritis.[11] Idiopathic causes have also been reported.[12,13]

Damage to the hypoglossal nerve produces characteristic clinical manifestations, mainly fasciculation, atrophy and deviation of the tongue on the affected side on protrusion, with dysarthria frequently reported. Hypoglossal nerve palsy is usually reported in combination with other cranial nerve symptoms such as facial palsy and ophthalmic manifestations. However, it is rare to see isolated hypoglossal nerve palsy and, when present, it is considered a diagnostic conundrum.

We report an unusual case of unilateral isolated hypoglossal nerve palsy. This case highlights the importance of interpreting subtle clinical signs and symptoms in the context of underlying pathology, and the role of the multidisciplinary team in the management of this case.

Case report

A 50-year-old Caucasian woman presented with an eight-week history of perceived 'thickening' of the left side of her tongue. At first she assumed it was related to a mouth ulcer, as she was prone to development of these. She reported that the left side of her tongue was tender to touch but otherwise she had no other symptoms. There was no impact on day-to-day activities such as swallowing or speech, nor did she recollect any significant events that might have corresponded to the onset of her symptoms.

Initially, she attended her general medical practice and was seen by numerous GPs in the practice, all of whom found the presentation quite peculiar. A second opinion was requested privately from an oral and maxillofacial surgeon, who referred her urgently to the Oral Medicine Department for further investigation.

Her medical history was unremarkable and she was not taking any medication other than occasional analgesics. She was a regular dental attender who had had no recent dental complaints or treatment. She was a non-smoker who drank one to two units of alcohol per week.

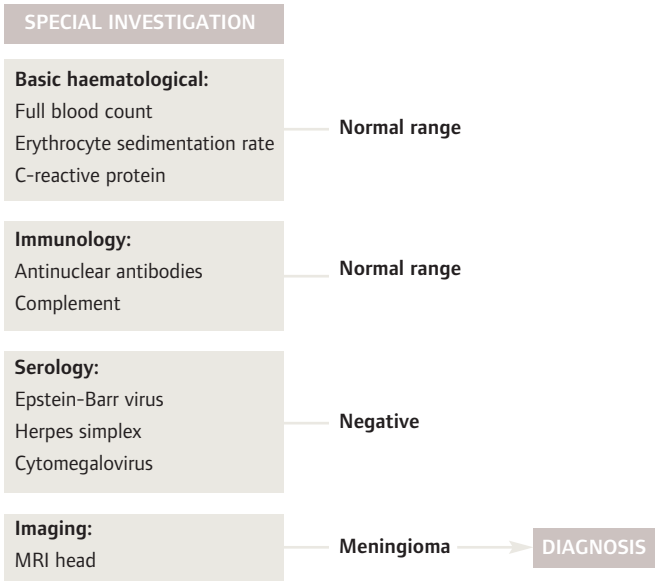

FIGURE 2: Special investigations to determine the cause of hypoglossal nerve palsy.

During extra-oral examination at the oral medicine clinic, there was no evidence of cervical lymphadenopathy, facial asymmetry or salivary gland swelling. Examination showed that cranial nerves I-XI were intact. On intra-oral examination she was partially dentate with a well-maintained dentition, good lubrication of all mucosal tissues and adequate oral hygiene. Examination of her tongue revealed mild wasting on the left side, which had a heaped appearance and quite marked tongue scalloping. There was visible fasciculation on the left lateral border and, on protrusion, the tongue deviated to the left side (**Figure 1**) with some limitation of tongue movements. There was no evidence of speckling, erythema or ulceration of soft tissues, nor any mass lesion palpated within the substance of the tongue.

The clinical impression was that of an isolated left hypoglossal nerve injury. A number of investigations were carried out in order to determine the cause of the suspected hypoglossal nerve injury (**Figure 2**). Haematological and biochemical investigations were undertaken, which revealed neutropenia, and this subsequently normalised on repeat testing. Immunological studies including complement, antinuclear antibody and double-stranded DNA were undertaken to identify an autoimmune aetiology. The results of these were negative. There were no signs of inflammation or infection, and no antibodies were identified for any infectious diseases.

In order to explore the possibility of a space-occupying lesion, an urgent magnetic resonance imagery (MRI) scan of the head was requested rather than a CT. This revealed a soft tissue lesion approximately 12x6mm in the left cerebellomedullary cistern, extending into the left hypoglossal canal (**Figure 3**). The case was discussed with a consultant neurosurgeon within the hospital trust who advised an urgent referral to their department and discussed the case at the next neuro-oncology multidisciplinary meeting. The general consensus based on imaging was that the lesion was likely to represent a meningioma. A conservative approach has been adopted due to the position of the lesion. The patient has regular review in neurology with repeat MRI imaging. One year on, there has been no significant change on imaging or in clinical presentation.

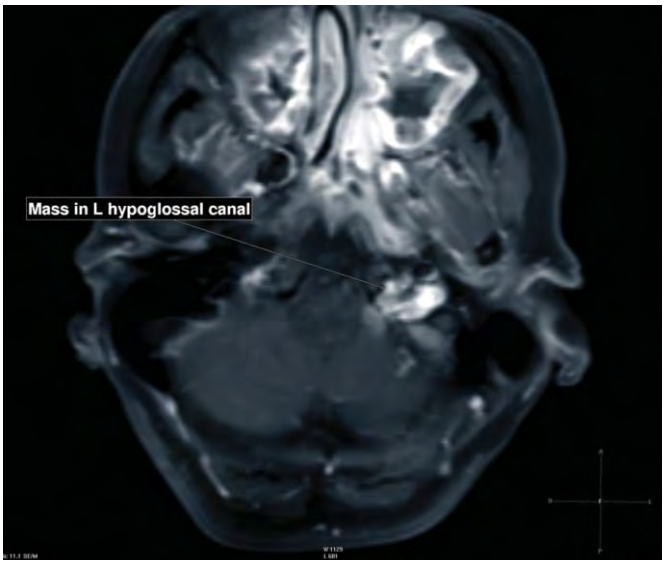

FIGURE 3: MRI T1 weight axial slice of posterior cranial fossa showing mass in left hypoglossal canal.

Discussion

The hypoglossal nerve is a motor nerve that innervates tongue musculature controlling its voluntary movements, and is also involved in speech and swallowing. Disorders affecting the function of this nerve lead to imbalanced action of the genioglossus muscles, causing the tongue to deviate towards the weakened side.

Hypoglossal nerve palsy is uncommon and rarely presents in isolation. Keane *et al*.,[4] in their case series of hypoglossal nerve palsy, found neoplastic aetiology in 85% of their cases, thus emphasising that isolated hypoglossal nerve palsy often suggests an ominous prognostic sign. However, a further case series of isolated hypoglossal nerve palsy[14] reported idiopathic hypoglossal nerve palsy in four out of nine cases and metastasis in three cases, with Arnold Chiari malformation and dural AV fistula as causes in the remaining cases.

The presentation of hypoglossal nerve palsy should raise suspicion of a sinister underlying pathology and should prompt referral for further investigations. In this case the hypoglossal nerve palsy presented as an isolated finding in the absence of any history or regional symptoms, which was in part reassuring.

Regardless of the suspected aetiology, haematological and autoantibody investigations in addition to imaging studies, including CT scan and MRI, are mandatory in the diagnostic approach to identify the causative pathology.[13] In our case blood investigations were normal, and the diagnosis of meningioma was made on the basis of MRI imaging and discussion at the neuro-oncology multidisciplinary team meeting.

Meningiomas are common neoplasms of the nervous system, arising from the meninges of the brain, and represent 19% of all primary intracranial tumours, of which approximately 90% are benign, and 2% are malignant.[15] However, meningiomas arising in the hypoglossal canal, are extremely rare and this is the fourth case to be reported in the literature so far.[15-17] The choice of treatment approach depends on several factors including: tumour type; size; compression of neural structures; patient age; symptoms; and, comorbid conditions. Should surgery be considered, there is currently no consensus regarding the ideal surgical approach for treating these lesions.[18]

Conclusion

We describe an unusual case of isolated hypoglossal nerve palsy, which has rarely been reported in the literature. This case emphasises the importance of detecting subtle changes in the oral cavity, the importance of special investigations in the diagnosis, and the role of the multidisciplinary team in managing the case.

Acknowledgement

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References

1. **Snell, R.S.** The cranial nerve nuclei and their central connection and distribution. In: **Snell, R.S.** *Clinical Neuroanatomy*. Wolters Kluer/Lippincott, Williams & Wilkins. Philadelphia; 2010; 11: 356.
2. **Chong, V.F., Fan, Y.F.** Hypoglossal nerve palsy in nasopharyngeal carcinoma. *European Radiology* 1998; 8 (6): 939-945.
3. **Greenberg, H.S., Deck, M.D.F., Vikram, B., Chu, F.C.H., Posner, J.B.** Metastasis to base of skull: clinical finding in 43 patients. *Neurology* 1981; 31: 530-537.

4. **Keane, J.R.** Twelfth nerve palsy. Analysis of 100 cases. *Arch Neurology* 1996; 53: 561-566.
5. **Theodotou, B., Mahaley, M.S.** Injury to the peripheral cranial nerve during carotid endarterectomy. *Stroke* 1985; 16: 894-895.
6. **Rubinsteine, M.K.** Cranial mononeuropathy as the first sign of intracranial metastasis. *Ann Intern Med* 1969; 70: 49-50.
7. **Mahadevappa, K., Chacko, T., Nair, A.** Unilateral hypoglossal nerve palsy due to vertebral artery dissection. *Clinical Medicine & Research* 2012; 10 (3): 127-130.
8. **Lindsay, F.W., Mullin, D., Keefe, M.A.** Subacute hypoglossal nerve paresis with internal carotid artery dissection. *Laryngoscope* 2003; 113 (9): 1530-1533.
9. **Morelli, R.J.** Intracranial neurilemomma of hypoglossal nerve: review and case report. *Neurology* 1966; 16: 709-713.
10. **Spitzer, C., Mull, M., Topper, R.** Isolated hypoglossal nerve palsy caused by carotid artery dissection. *J Neurol* 2001; 248: 909-910.
11. **Al Ramzi, A., Kassem, M., Barghash, A.** Temporary unilateral hypoglossal nerve palsy secondary to infectious mononucleosis: A case report. *Future Dental Journal* 2016; 2: 1-3.
12. **Freedman, M., Jayasundara, H., Stassen, L.** Idiopathic isolated unilateral hypoglossal nerve palsy: a diagnosis of exclusion. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology and Endodontology* 2008; 106: 22-26.
13. **Ho, M., *et al.*** Persistent idiopathic unilateral isolated hypoglossal nerve palsy: a case report. *British Dental Journal* 2004; 196: 205-207.
14. **Combarros, O., Alvarez de Arcaya, A., Berciano, J.** Isolated unilateral hypoglossal nerve palsy: nine cases. *J Neurol* 1998; 245: 98-100.
15. **Zulkiflee, A.B., Prepageran, N., Rahmat, O., Jayalaskhmi, P., Sharizal, T**. Hypoglossal nerve tumor: a rare primary extracranial meningioma of the neck. *ENTJ* 2012; 91: E26-E29.
16. **Neeff, M., Baysal, E., Homer, J., Gillespie, J., Ramsden, R**. Intracranial/extracranial meningioma arising in the hypoglossal canal: case report. *Skull Base* 2007; 17: 325-330.
17. **Calzada, G., Isaacson, B., Yoshor, D., Oghalai, J.S**. Surgical approaches to the hypoglossal canal. *Skull Base* 2007; 17: 187-196.
18. **Tatagiba, M., Koerbel, A., Roser, F**. The midline suboccipital subtonsillar approach to the hypoglossal canal: surgical anatomy and clinical application. *Acta Neurochir* 2006; 148: 965-969.

CPD questions

To claim CPD points, go to the MEMBERS' SECTION of www.dentist.ie and answer the following questions:

1. The hypoglossal nerve innervates all muscles of the tongue except:

- ○ A: Hypoglossus muscle
- ○ B: Palatoglossus muscle
- ○ C: Styloglossus muscle
- ○ D: Genioglossus muscle
- ○ E: Superior longitudinal muscle

2. Which of the following statements about hypoglossal nerve palsy is false?

- ○ A: In unilateral palsy, the tongue should deviate away from the weakened side
- ○ B: Damage to the hypoglossal nerve may not always be noticeable to patients
- ○ C: In unilateral palsy, the tongue should deviate towards the weakened side
- ○ D: It can present with atrophy and fasciculations of the tongue
- ○ E: It can present with facial palsy and ophthalmic manifestations

3. **The percentage of intracranial meningiomas that are benign is approximately:**

- ○ A: 10%
- ○ B: 30%
- ○ C: 50%
- ○ D: 70%
- ○ E: 90%

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