IN BRIEF

- XII nerve palsy can be easily detected on routine oral examination.
- Clinical signs — deviation of the tongue, muscle wasting and muscle fasciculations.
- Malignancy or cerebrovascular accident could cause XII nerve palsy.

VERIFIABLE
CPD PAPER

Persistent idiopathic unilateral isolated hypoglossal nerve palsy: a case report

M. W. S. Ho¹, M. J. Fardy² and St J. V. Crean³

Isolated hypoglossal nerve palsy (IHNP) although a rare condition, has been previously reported. A literature review revealed that in most cases, IHNP indicates the presence of an intracranial or extracranial space occupying lesion, head and neck injury, vascular abnormality, infection, autoimmune disease or neuropathy. Reports of idiopathic cases are rare and the vast majority of IHNP are reversible. We report a case of persistent idiopathic unilateral hypoglossal nerve palsy, with an emphasis on the investigations necessary to be undertaken on presentation of such a lesion.

Idiopathic isolated hypoglossal nerve palsy (IHNP) has been documented by several authors. A search through the literature of the past decade has revealed that only one case of persistent idiopathic IHNP has been reported.¹ Almost all of the reported cases of idiopathic IHNP, have been self-limiting.^{2–5} Lee *et al.*² believe that self-limiting idiopathic hypoglossal nerve mimics Bell's palsy of the VIIth cranial nerve.

CASE REPORT

A 25-year-old Caucasian female presented with deviation of her tongue, towards the right hand side. She had first noticed the problem 5 years previously, after she had pulled a muscle in her neck. A visit to her general medical practitioner failed to provide a diagnosis. As a school teacher,

she had experienced difficulty speaking and found reading aloud for long periods a big problem. There was also an increased awareness of the problem when she was running or rowing. She also noticed some difficulty swallowing. She began seeing a speech therapist on a regular basis, in an attempt to improve her tongue movements.

About 3 years ago, she was referred to the Department of Neurology for investigation of the problem. She underwent a range of haematological investigations: full blood count, urea and electrolytes, glucose, liver function test, erythrocyte sedimentation rate, thyroid function test, vitamin B₁₂, folate, serum electrophoresis, auto-

antibodies and serology for treponema, all of which were unremarkable.

A chest x-ray (CXR) was normal, with no sign of tuberculosis or sarcoidosis. Computerised tomography (CT) was reported as normal with special note made of the normal appearance of the carotid canals. Magnetic resonance imaging (MRI) was equally unremarkable with no parenchymal signal abnormality in either the brainstem or the cerebellar hemispheres. Cerebro-spinal fluid (CSF) showed slight elevation of protein levels but abnormal cells were absent.

She was next seen by the ear, nose and throat team, who after examination were unable to ascertain the cause of the tongue

¹Senior House Officer in Oral and Maxillofacial Surgery

²Consultant in Oral and Maxillofacial/Head and Neck

Surgery, ³Senior Lecturer and Honorary Consultant in Oral and Maxillofacial Surgery, Department of Oral Surgery, Oral Medicine and Oral Pathology, University of Wales College of Medicine, Cardiff

Correspondence to: M. W. S. Ho, 65 Youghal Close,

Pontprennau, Cardiff CF23 8RN

Email: michaelws_ho@hotmail.com

Refereed Paper

DOI: 10.1038/sj.bdj.4810980

Received 19.02.03; Accepted 20.06.03

© British Dental Journal 2004; 196: 205–207

Fig.1 Photograph of the patient's tongue deviating to the right when protruded

Table 1 Possible causes of hypoglossal nerve palsy

Metastatic disease at base of skull
Medical causes eg sarcoidosis, autoimmune disease, vasculitis
Arnold Chiari malformation
Dural arteriovenous fistula of the transverse sinus
Periostitis of hypoglossal canal
Post-retropharyngeal infection
Surgical procedure near neck
Acute poliomyelitis
Syringobulbia
Thrombosis of median branches of vertebral artery
Multiple sclerosis
Carotid artery dissection/aneurysm
Diabetes mellitus
Lacunar infarct over the hypoglossal nerve nucleus
Rare sequelae of central venous catheterisation ⁶
Head and neck trauma ⁷ eg subluxation of odontoid process
Fracture through the occipital condyle ⁸
Glomus tumour
Meningioma

deviation. A referral to the Department of Oral and Maxillofacial Surgery followed, to exclude any local structural disease (though this was highly unlikely, in the absence of progression or pain).

Her medical history was unremarkable. There was no family history of idiopathic IHPN or any other neurological disorders. She had stopped smoking 3 years previously and drank approximately 6 units of alcohol per week.

Clinical examination

The patient looked well. There was no cervical lymphadenopathy, facial asymmetry and no external scars of the neck or scalp. Cranial nerves I–XI were intact generally except for some decreased sensation in the distribution of the right lingual nerve. Left and right fundoscopy was normal. Examination of hypoglossal nerve function revealed fasciculation, and muscle atrophy on the right side of the tongue. The tongue deviated to the right side as the patient opened her mouth (Fig. 1). The muscles of mastication displayed good function. Examination of the upper and lower limbs bilaterally for tone, power, reflexes, sensation and co-ordination was grossly normal.

An orthopantomogram (OPT) revealed nothing significant apart from a carious lesion in tooth LL8 (38) which was brought to the attention of the patient.

Unilateral isolated hypoglossal nerve palsy suggested that the cause might be a local vasculitic lesion or a cerebrovascular accident. However, there was no evidence to support these provisional diagnoses.

She was last seen in the clinic early this year as part of her follow-up. The deviation of the tongue to the right

remained unchanged. The patient also felt that the situation had remained stable. The fasciculation of the tongue still bothered her as she was a teacher but swallowing was no longer a problem. However, she experiences occasional gagging sensation. Clinically, marked

deviation of the tongue to the right side with muscle fasciculation was evident. Speech and language therapy assessment was arranged for the patient for further exercise sessions if required

DISCUSSION

The presentation of hypoglossal nerve palsy could be the harbinger of a more sinister underlying pathology. Clinicians should be suspicious in such circumstances and be prepared to follow an investigative pathway designed to reveal a possible diagnosis. In this case a XIIth nerve palsy presented in the absence of any obvious history. The first step for the attending clinician is to consider the possible causes of isolated HNP. The potential causes are outlined in Table 1.

A careful history is essential when considering these possibilities. The absence of any regional symptoms and negative history in our patient ruled out the presence of metastatic disease at the skull base, post-retropharyngeal infection and surgical procedure in the neck. In Arnold-Chiari malformation, the cerebellar tonsils and medulla are malformed and herniate through the foramen magnum causing progressive hydrocephalus with mental retardation, optic atrophy and ocular palsies, and spastic paralysis of the limbs.

Fig. 2 Potential pathway of investigations in a patient presenting with isolated hypoglossal nerve palsy

In dural arteriovenous fistula of the transverse sinus the patient may have hypoglossal palsy resulting from ischaemia in the region of the hypoglossal nucleus, as would thrombosis to the median branches of the vertebral artery. MRI and CT studies would aid in the diagnosis of the two latter conditions and also reveal any carotid artery dissection/aneurysm or periostitis of hypoglossal canal (where narrowing of the hypoglossal canal would be evident). In syringobulbia, patients may present with wasting and weakness of the hands and arms with loss of pain and temperature sensation in a cape distribution. None of the above signs and symptoms were detected in our patient. Diabetes was excluded by random blood glucose and there was no history of trauma. The patient had not received any intravenous treatment and the CT and MRI did not demonstrate any intracranial tumour.

This wide group of possible diagnoses, may only reveal themselves after a number of investigations. One suggested pathway of investigation is that followed in this case, and serves as a possible investigative approach when faced with such a diagnostic problem. It is outlined in Figure 2

Thus with a diagnosis of exclusion the clinician can make a true diagnosis of idiopathic IHNP. Should such a case present to the general dental practitioner, referral to the local neurology or maxillofacial unit is advised where the investigations in Figure 2 can be undertaken. In the situation where a cerebrovascular accident is suspected and this cannot be ruled out by the

GDP, an urgent referral to an acute medical receiving/stroke unit is required. The investigations can be carried out at appropriate times thereafter.

One other case of unresolved idiopathic IHNP has been reported and has a number of similarities to our case.¹ Bagan Sebastian *et al.* reported a case of a 24-year-old woman, who for the previous 10 years had noted alterations to the motility of her tongue and displacement to the right on protrusion. She also had slight dysarthria. Neurological examination was normal apart from the right hypoglossal nerve palsy. As with our case, the absence of any other signs and symptoms excluded the causes listed in Table 1. Basic and special haematological investigations, radiological investigations and infectious agents serology were also negative and the aetiology of the hypoglossal nerve palsy remained unknown. The patient was followed up 12 months, when she claimed to have adapted to the situation and reported no problems in swallowing, chewing or speaking.

In our case, the pathway illustrated in Figure 2 was followed and all the findings were negative apart from a slightly elevated protein level in the CSF. Thus the diagnosis of persistent idiopathic unilateral IHNP was made. The isolated hypoglossal nerve palsy in our patient has persisted for the past 5 years. At the most recent review, no other neurological problem was evident. We have recommended regular review as a demyelinating disease remains a possibility,

thorough neurological investigation and a repeat of the MRI scan every 3–5 years in the absence of any other neurological symptoms.

A patient with hypoglossal nerve palsy could present in the dental surgery to a dental healthcare worker and one should be aware of the significance of its oral manifestation. The likely differential diagnoses should be born in mind and appropriate referral should be arranged while first line investigations are carried out where possible.

The authors would like to thank Professor C M Wiles, Department of Neurology, University Hospital of Wales for referring this patient to our department and allowing us to report this case

1. Bagan-Sebastian J V, Milian-Masanet M A, Penarrocha-Diago M *et al.* Persistent idiopathic unilateral hypoglossal nerve palsy. *J Oral Maxillofac Surg* 1998; **56**: 507–510.
2. Lee S S, Wang S J, Fuh J L *et al.* Transient unilateral hypoglossal nerve palsy: a case report. *Clin Neurol Neurosurg* 1994; **96**: 148–151.
3. Sugama S, Matsunaga T, Ito F *et al.* Transient, unilateral, isolated hypoglossal nerve palsy. *Brain Dev* 1992; **14**: 122–123.
4. Pandey V P, Deshpande R, Talati R *et al.* Isolated idiopathic unilateral XIIth nerve palsy. *J Assoc Physicians India* 2001; **49**: 293–294.
5. Giufridda S, Lo Bartolo M L, Nicoletti A *et al.* Isolated, unilateral, reversible palsy of the hypoglossal nerve. *Eur J Neurol* 2000; **7**: 347–349.
6. Isolated palsy of XIIth nerve after central venous catheterisation. *Br Med J* 1984; 1042.
7. Omura S, Nakajima Y, Kobayashi S *et al.* Oral manifestation and differential diagnosis of isolated hypoglossal nerve palsy: report of two cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1997 Dec; **84**: 635–640.
8. Castling B, Hicks K. Traumatic isolated unilateral hypoglossal nerve palsy – case report and review of the literature. *Br J Oral Maxillofac Surg* 1995; **33**: 171–173.