Persistent isolated unilateral hypoglossal nerve palsy

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Abstract

Idiopathic isolated unilateral hypoglossal nerve palsy is a rarely observed finding in clinical practice. Cases of persistent idiopathic isolated hypoglossal nerve palsy are extremely rare, one similar case documenting only one other documented case in the literature. Cases of this finding in patients with epilepsy further make this clinical finding a very rare entity. The presenting case is a 22 year old who complains of deviation of the tongue to the left side upon protrusion and atrophy of the left side of the tongue. The patient was diagnosed with epilepsy at age 10 and had been successfully controlled with medication remaining seizure free for over 10 years.

Introduction

The hypoglossal nerve plays an important role in speech and swallowing. Despite the nerves important function, damage to the nerve does not usually present with any functional loss or symptoms that would be overtly noticeable to most patients [1,2]. Patients may become aware of such lesions when brushing their teeth or when visiting a dentist. Causes of hypoglossal nerve palsy include intracranial space occupying lesions (49%), trauma (12%), stroke (6%), hysteria (6%), surgery (5%), multiple sclerosis (5%), infection (4%), Guillain-Bare syndrome (4%) and idiopathic causes (3%) [3]. The patient in this case has persistent isolated unilateral left sided hypoglossal nerve palsy and cannot recall when the symptoms first started. The patient continues to experience the palsy and has not had the symptoms resolve or change. The patient has not experienced any challenges because of the symptoms.

Case report

A 22-year-old patient was seen by a neurologist for a chief complaint of left sided tongue deviation upon protrusion. Fasciculations were also present on protrusion. There is also atrophy of the tongue on the left side. CN1-XI are intact, strength was 5/5 in all muscle groups, sensation to sharp and dull were intact, DTR’s were 2+ bilaterally, Babinski sign was negative, cerebellum exam was normal with a negative Romberg sign and the patient has a normal gait. The patient has no constitutional findings and stable vital signs.

The patient was diagnosed with epilepsy at age 10. At the time the patient experienced a total of 4 tonic-clonic seizures, and an unknown number of absence seizures. The patient was subsequently treated with valproic acid and has been seizure free for over 10 years. The patient has no history of head trauma, paralysis, paresis, paresthesia, weakness, and has no focal neurological signs. The patient has no history of meningitis or other infectious sequelae.

Multiplanar multisquence MR images were obtained of the skull base with and without gadolinium contrast on a 1.5 Tesla closed MR scanner (Figures 1-3).

The patient has no constitutional findings and stable vital signs.

Discussion

The hypoglossal nerve is the motor nerve responsible for movement of the tongue muscles playing an important role in articulation of speech [4]. Isolated hypoglossal nerve damage presents less frequently than isolated damage to cranial nerves III, IV, VI, and is a rare finding in clinical practice [5]. The motor distribution of the hypoglossal nerve is highly complex. The hypoglossal nerve has 4 associated sub nuclear components [6]. The hypoglossal nerve is commonly used in transplant surgery for facial paralysis, suggesting damage to the hypoglossal nerve may present in subtle ways [6]. The hypoglossal nerve can be damaged anywhere from the medullary nucleus, hypoglossal canal or skull base [1]. Although the relative benign presentation of hypoglossal nerve palsy and its limited consequences for function, proper assessment and evaluation is necessary to rule out serious underlying causes of this rare pathology.

With contrast no abnormal enhancement was seen within the skull base.

Ho et al. [2] identified a number of possible causes of hypoglossal nerve palsy. The most common causes are trauma, surgery, infection, stroke, and medical conditions such as Guillain-Bare syndrome and multiple sclerosis. Other causes include idiopathic conditions, such as idiopathic hypoglossal palsy. In rare cases, idiopathic hypoglossal palsy may be associated with other neurological conditions, such as stroke or multiple sclerosis. The patient in this case had a history of epilepsy and was seizure free for over 10 years. The patient had no other history of neurological conditions.

Ho et al. [2] described the clinical presentation of idiopathic hypoglossal palsy as deviation of the tongue to the side of the palsy, atrophy of the tongue, and fasciculations. The patient in this case had deviation of the tongue to the left side, atrophy of the left side of the tongue, and fasciculations were noted on protrusion.

The hypoglossal nerve is important for speech and swallowing. The hypoglossal nerve is the motor nerve responsible for movement of the tongue muscles playing an important role in articulation of speech [4]. Isolated hypoglossal nerve damage presents less frequently than isolated damage to other cranial nerves. The hypoglossal nerve has a highly complex motor distribution with 4 associated sub nuclear components. The hypoglossal nerve is commonly used in transplant surgery for facial paralysis, suggesting damage to the hypoglossal nerve may present in subtle ways. The hypoglossal nerve can be damaged anywhere from the medullary nucleus, hypoglossal canal or skull base. Although the relative benign presentation of hypoglossal nerve palsy and its limited consequences for function, proper assessment and evaluation is necessary to rule out serious underlying causes of this rare pathology.

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nerve palsy including; metastatic disease at the base of the skull, sarcoidosis, autoimmune disease, vasculitis, Arnold Chiari malformation, dural AV fistula of the transverse sinus, periostitis of the hypoglossal canal, post-arthroplasty infection, surgical procedure near neck, acute polymyelitis, syringobulbia, thrombosis of the median branches of the vertebral artery, multiple sclerosis, carotid artery dissection or aneurysm, diabetes mellitus, lacunar infarct over the hypoglossal nucleus, complication of central venous catheterization, head and neck trauma, fracture through the occipital condyle, glomus tumor and meningioma.

Conclusion

This unique case of a patient who suffers from persistent isolated hypoglossal nerve palsy with epilepsy as a co-morbidity presents the case with a unique perspective. The clinical finding of persistent isolated hypoglossal nerve palsy itself is a rare clinical entity; made even more rare when this finding occurs in a patient with epilepsy. Further exploration into epilepsy as a possible cause of isolated hypoglossal nerve palsy should be made.

References

1. Ahmed SV, Akram MS (2014) Isolated unilateral idiopathic transient hypoglossal nerve palsy. BMJ Case Rep. [Crossref]
2. Ho MWS, Fardy MJ, Crean SJV (2004) Persistent idiopathic unilateral isolated hypoglossal nerve palsy: A case report. Br Dent J 196: 205-207. [Crossref]
3. Keane JR (1996) Twelfth-Nerve PalsyAnalysis of 100 Cases. Arch Neurol 53: 561-566. [Crossref]
4. Mahadevappa K, Chacko T, Nair AK. Isolated unilateral hypoglossal nerve palsy due to vertebral artery dissection. Clin Med Res 10: 127-30. [Crossref]
5. Shikino K, Noda K, Ikusaka M (2017) Transient Idiopathic isolated unilateral Hypoglossal nerve palsy. J Gen Intern Med 28: 591. [Crossref]
6. Yoon JH, Cho KL, Lee HJ, Choi SH, Lee KY, et al. (2011) A case of idiopathic isolated hypoglossal nerve palsy in a Korean child. Korean J Pediatr 54: 515-517. [Crossref]