

Isolated unilateral hypoglossal nerve palsy due to schwannoma

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Abstract

As benign nerve sheath tumors, schwannomas are located mostly in the head and neck region, but rarely originate from the hypoglossal nerve. A woman aged 53 years presented with non-progressive dysarthria and unilateral tongue atrophy, which she had had for 2 years. Neurologic examination revealed left-sided, isolated hypoglossal nerve palsy. Chronic axonal neuropathy of the left hypoglossal nerve was seen in electromyographic (EMG) study. With cranial magnetic resonance (MR) imaging, a mass lesion in the left cerebellomedullary cistern was detected. A biopsy was not possible due to its critical location. Due to the non-progressive clinical course and radiologic features of the mass, it was considered as schwannoma. Six-monthly follow-up with MR imaging and EMG was performed three times, and there was no change in the findings. We aimed to provide clinical, radiologic and electrophysiologic findings of isolated unilateral hypoglossal nerve palsy due to schwannoma.

Keywords: Hypoglossal nerve, schwannoma, isolated, unilateral, hypoglossal nerve palsy

INTRODUCTION

The hypoglossal nerve is a purely motor cranial nerve (CN) that originates from the brainstem and goes through the skull base to the its target organ, the tongue (1). Hypoglossal nerve palsy usually presents with other signs because of its close proximity to other brainstem or cervical structures.

CASE PRESENTATION

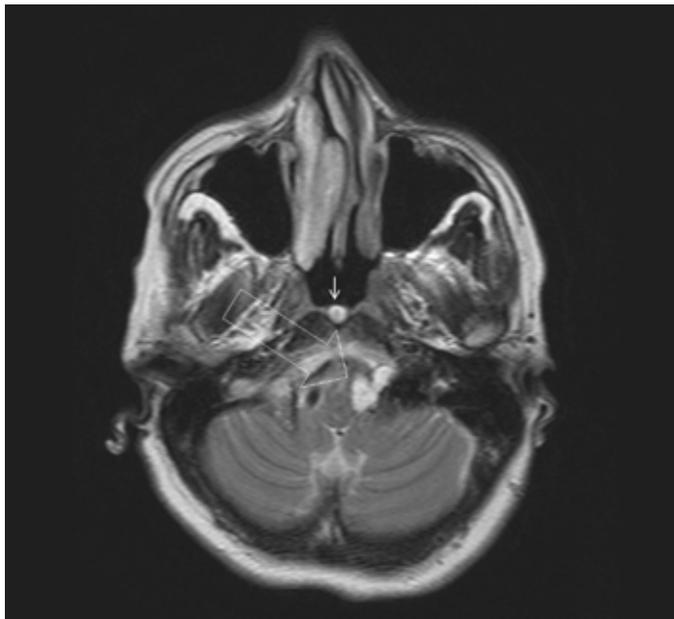
A woman aged 53 years presented with tongue weakness and impediment of speech. The symptoms began 2 years ago with pain in the left periauricular region, followed by tongue debility and impediment of speech. The patient had been given gabapentin for her symptoms by another center before presenting to us. After medication with gabapentin, her pain improved dramatically; however, the tongue debility remained. Physical examination showed that speech was slightly dysarthric and the left side of the tongue was noticeably atrophic. The tongue tended to point rightward in the mouth, and to leftward out of the mouth. No other pathology was noted in examination.

The patient had no known systemic disease, and there was no history of trauma or operation temporally related to disease onset. In the neurologic examination, isolated hypoglossal nerve palsy and apparent tongue atrophy were found. Magnetic resonance (MR) imaging depicted a mass lesion with a cystic component in the left cerebellomedullary cistern (Figure 1). The mass indented the bulbous and followed the route of the left CN XII. The mass showed peripheral contrast enhancement. The findings suggested a schwannoma of the CN XII (Figure 2). Cranial and cervical MR angiography confirmed that vascular structures were intact. Cranial computed tomography (CT) scan revealed no destruction to bony structures. Electromyography (EMG) study showed chronic neurogenic changes in the tongue muscles, with no abnormalities related to the other lower cranial nerves. Surgery and biopsy were avoided because, with the exception of the tongue atrophy, the symptoms were nonprogressive; the hypoglossal nerve involvement was clinically and electrophysiologically isolated; the lesion had a benign nature on the imaging studies; and there was a high complication risk due to location in a surgically difficult region. Rather, elec-

Table 1. Segmental approach to the etiologic causes of hypoglossal nerve palsy (5, 8, 12)

Medullary segment	Cisternal segment	Skull base segment	Extracranial segment
*Glioma	*Vertebrobasilar aneurysm/dolicoectasis	*Metastasis	*Carcinoma
*Infarction	*Rheumatoid arthritis	*Nasopharyngeal carcinoma	*Lymphoma
*Hemorrhage	*Meningioma	*Glomus tumors	*Soft tissue sarcoma
*Demyelination	*Cordoma	*Nerve sheath tumors	*Salivary gland tumors
*ALS	*Basal meningitis	*Skull base fractures	*Metastasis
*Syringobulbia	*Subarachnoid hemorrhage	*Primary bone tumors	*Infections
*Poliomyelitis	*Arnold-Chiari malformation	*Fibrous dysplasia	*Carotid artery dissection/aneurysm
	*Nerve sheath tumors	*Paget disease	*Trauma
		*Skull base infection	*Endarterectomy
			*Radiation
			*Jugular vein thrombosis

Figure 1. Transverse T2-weighted MR imaging through the bulbous shows a hyperintense mass (arrow) in the left cerebellomedullary cistern. The mass indents the bulbous and follows the route of the left cranial nerve (CN) XII. Note a coincidental Thornwald cyst in the nasopharyngeal mucosa (small arrow).

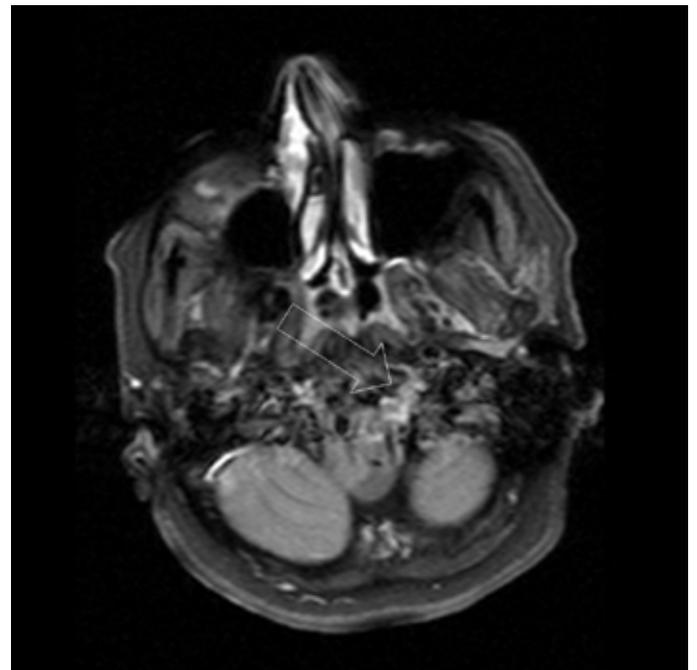


trophysiologic and radiologic follow-up was scheduled. The patient was clinically stable, and there had been no change in the findings of EMG studies or MR imaging, which had been performed three times with six-months apart. For this report, written informed consent was obtained from the patient.

DISCUSSION

Hypoglossal nerve fibers arise from the hypoglossal nucleus in the medulla. Fibres leave the cranium through the hypoglossal canal as the hypoglossal nerve (1). It moves toward the

Figure 2. Transverse fat saturated postcontrast T1-weighted magnetic resonance (MR) imaging shows prominent peripheral enhancement.



neck close to the internal carotid artery, jugular vein, and vagal nerve, and reaches the intrinsic and extrinsic tongue muscles. The hypoglossal nerve is separated anatomically into the intraaxial, cisternal, skull base, and extracranial segments (2).

Hypoglossal nerve palsy is frequently seen with symptoms related with brainstem and lower cranial nerves (3, 4). Diseases that appear with hypoglossal nerve palsy are presented by segmental location in Table 1. In the 100-case series of Keane, the most prevalent cause of hypoglossal nerve palsy was speci-

fied as tumor and trauma (5). Most of the cases were unilateral, accompanying cranial neuropathy and brainstem symptoms. However, their article did not specify the rate of isolated XIIth nerve palsy. In Combarros et al.'s nine patients with unilateral hypoglossal nerve palsy, tumor was determined in three patients (6). In the other cases, Chiari malformation and transverse sinus dural arteriovenous fistulae were determined as etiologic causes (6). Boban et al. reported four isolated hypoglossal nerve palsies with different etiologies. Parenchymal involvement due to acute demyelinating encephalomyelitis, tumor metastasis, and internal carotid pseudoaneurysm were found as underlying causes (3). In another three unilateral isolated hypoglossal nerve palsy cases reported by Mateen et al. no etiologic cause could be identified (7).

Schwannoma, as a slowly growing and mostly benign tumor of schwann cells, is found in the head and neck region with an occurrence of 25-45%. It can originate from cranial and other peripheral nerves, sympathetic chain, thyroid gland, larynx, posterior pharyngeal wall, and other structures (8-11). Intracranial schwannoma mostly arises from the vestibular nerve (12). The hypoglossal nerve is affected in only 5% of cases (13). In Keane's case series, the most frequently seen etiologic cause was reported as tumors, but the percentage of schwannoma among them was not specified (5). Piccirilli et al. published a review of 105 patients with hypoglossal nerve palsy. It was found that schwannoma usually originated from the intracranial segment of the nerve (14). It was stated that other structures could be affected dependent on the direction and also extension of the tumor, and isolated hypoglossal nerve palsy was specific for extracranial neurinoma in the 90% of cases.

CONCLUSION

Isolated hypoglossal nerve palsy is rarely seen, but it has various etiologic causes. Clinical examination findings taken with the segmental location of the nerve would help to identify diagnostic tools for correct diagnosis among probable causes.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was obtained from patient who presented in this case report.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - N.D., İ.Ö., S.M.; Design - İ.Ö., N.D.; Supervision - İ.Ö.; Resources - N.D., İ.Ö., S.M.; Materials - İ.Ö., N.D., S.M.;

Data Collection and/or Processing - İ.Ö., N.D., S.M.; Analysis and/or Interpretation - İ.Ö., N.D., S.M.; Literature Search - N.D.; Writing Manuscript - İ.Ö., N.D., S.M.; Critical Review - İ.Ö., N.D., S.M.

Conflict of Interest: Authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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