

CASE REPORT

OLGU SUNUMU

UNILATERAL ISOLATED HYPOGLOSSAL NERVE PARALYSIS WITH UNUSUAL CAUSES

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ABSTRACT

We herein describe two rare cases of unilateral hypoglossal nerve paralysis due to unusual causes. Two of the cases presented with progressive dysphagia and speech impairment. All laboratory investigation were unremarkable except radiologic finding related with the lesion which is pressed on the nerve. Diagnoses of the lesions were established with biopsy at the first patient and digital subtraction angiography (DSA).

Keywords: Hypoglossal nerve paralysis, chordoma, aneurysm.

İZOLE UNİLATERAL HİPOGLOSSAL SİNİR FELCİNİN NADİR NEDENLERİ

ÖZ

Burada, nadir nedenlerden dolayı ortaya çıkmış unilateral hipoglossal sinir felci vakalarını sunduk. Olguların ikisi de progresif disfaji ve konuşma bozukluğu ile kliniğimize başvurdu. Radyolojik bulgular dışında sinire bası yapan lezyonun diğer laboratuvar incelemelerinde herhangi bir özellik yoktu. Tanı bir hastada biyopsiyle, diğer hastada ise dijital subtraksiyon anjiyografi (DSA) ile kondu.

Anahtar Sözcükler: Hipoglossal sinir felci, kordoma, anevrizma.

INTRODUCTION

Hypoglossal nerve palsy is a rare sign of neurologic abnormalities rather than symptoms. Clinical manifestations are unilateral atrophy of the tongue, speech difficulty and may have difficulty of swallowing. It is most commonly caused by tumours (49%), trauma (12%), strokes (6%), surgery (5%), and infection (5%) respectively and only 3% of cases are diagnosed as idiopathic (1).

We report two adult patients with unilateral hypoglossal nerve palsy, caused by unusual etiologies, and describe the investigations used in diagnosis.

CASE REPORTS

Case I

MO with an age of 30 years old man was referred to the outpatient clinic with a 2-months history of dysphagia, speech impairment and complaint of sustained unilateral head pain localized on left occipital base. His medical history was unremarkable. There was no history of trauma, infection or cerebrovascular accident, and to our knowledge he did not take any drugs regularly. Only he had a jaw implantation 7 years ago with no complication.

On examination he had no facial asymmetry,

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but had mild dysarthria and found it difficult to articulate certain words. Intra-orally there was wasting of the left side of the tongue with atrophy, and on protrusion it deviated to the affected side (Figure I). The oral cavity was otherwise normal and neurological examination showed no abnormality.



Figure I. Tongue is deviated to left side on protrusion and it is on the same side.

Case II

DA, 59 years old woman was referred with a 6 months history of dysphagia and speech impairment. She has a medical history of atopic eczema for 15 years. There was no history of trauma, infection, or cerebrovascular accident, and to our knowledge she did not take any drugs regularly.

On examination she had no facial asymmetry, but had mild dysarthria and found difficult to articulate certain words. Intra-orally there was wasting of the right side of the tongue with atrophy, fasciculation and on protrusion it deviated to the affected side (Figure II). Neurological examination showed no abnormality then that.



Figure II. Deviation of the tongue to the right side on protrusion and it is atrophied on the same side.

Investigation: Both patients had a range of hematological and biochemical investigations which were all within normal ranges, including his clotting profile. Immunological studies (rheumatoid factor, complement, antinuclear and anti-DNA antibodies) were normal. Angiotensin-converting enzyme (ACE) was in normal range. On the other hand, the infection markers were all negative (brucella, salmonella, syphilis, borrelia burgdorferi, anti tuberculosis Ig M and Ig G). Marker for malignant tumours were all in normal ranges (Carcinoembryonic antigen (CEA), prostate-specific antigen (PSA), free PSA, carbohydrate antigen (CA) 19-9, CA 125, alpha fetoprotein (α FP), CA 72-4).

In the second case a needle electromyography (EMG) of the tongue showed abnormal spontaneous activities, such as positive sharp waves and fibrillation potential with polyphasic motor unit potentials and reduced recruitment in the right genioglossus muscle. On the other hand, needle EMG of upper and lower extremities muscles were normal, motor neuron disease excluded via neurologic and EMG finding.

The normal chest radiograph excluded sarcoidosis and tuberculosis. Computed tomography of thorax, upper and pelvic abdomen imaging showed no disease.

Other investigations: In the first case; whole cervical and vertebral computed tomography with angiography showed left occipital hypoglossal condyle bone was destructed with 4x 3 cm mass.

Posterior fossa magnetic resonance imaging (MRI) revealed a heterogeneous mass with contrast originate from inferior clivus and protracted to cervical 1 (C1) prevertebral space of left occipital region (Figure III, IV). There was no evidence of intracranial aneurysm or arteriovenous malformation.

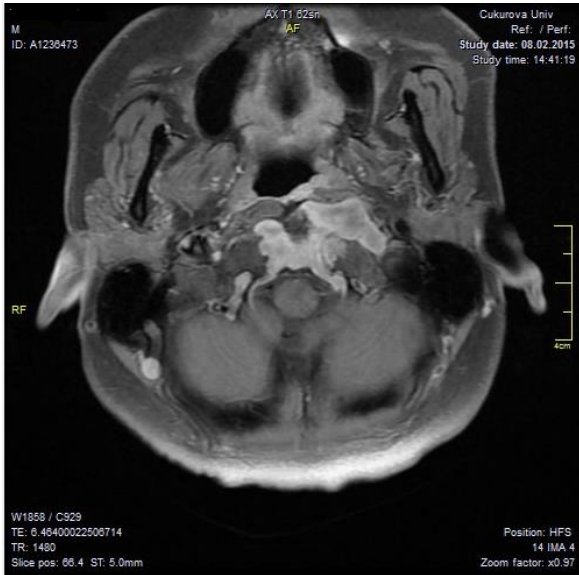


Figure III. Posterior fossa MRI: Left occipital hypoglossal condyle bone was destroyed with 4x 3 cm mass.

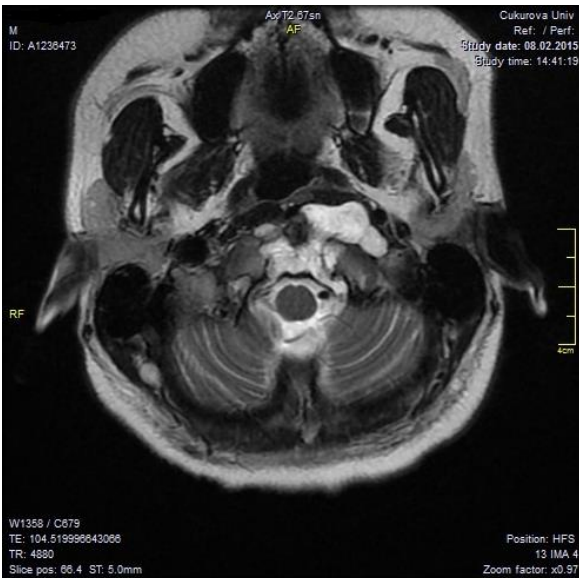


Figure IV. Posterior fossa MRI: Left occipital hypoglossal condyle bone was destroyed with 4x 3 cm mass.

After this result the patients underwent to the Tc- MDP whole-body bone scanning. In this

investigation there was no osteolytic bone metastases and normal finding were reported.

There were no evidence of infection, traumatic or vascular structural injury other than extra cranial lesion, he was diagnosed with localized occipital condyle bone lesion assault to the hypoglossal nerve. He was underwent of biopsy and then surgery. Microscopic examination of the biopsy specimen epithelioid tumor cells in cords and islands suspended in a mucinous myxoid background. The epithelioid cells are slightly elongated, nucleus are round and uniform.

Tumor cells were positive for EMA, immunohistochemically, cytokeratin and GFAP, S100 were negative (Figure V).

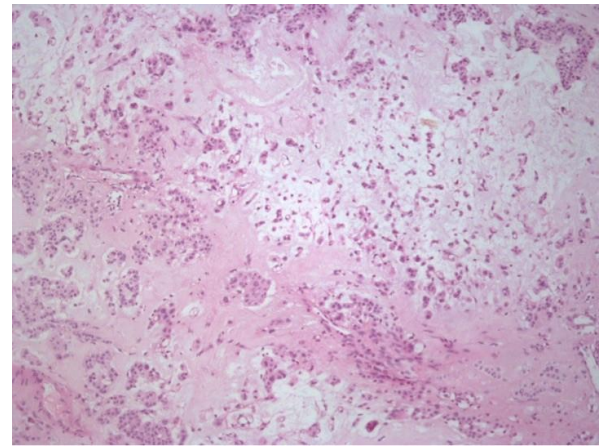


Figure V. Epithelioid tumor cells in cords and islands suspended in a mucinous myxoid background (H&E x 100).

In the second case; cerebral and posterior fossa magnetic resonance imaging (MRI) showed no ischemic lesion only revealed cerebral cortical atrophy. Cervical and vertebral magnetic resonance angiography showed right internal carotid artery aneurysm. After that result, digital subtraction angiography (DSA) achieved and saccular aneurysm on petrosal segment of the right internal carotid artery (ICA) have been shown (Figure VI, VII).

Informed consent was filled out by the patients for this paper.

DISCUSSION AND CONCLUSION

Hypoglossal nerve which is a pure motor nerve that innervating intrinsic as well as extrinsic tongue muscle. Hypoglossal nerve injury (HNI) is not an uncommon finding. Especially Keanne and et al. reported 100 cases diagnosed HNI with

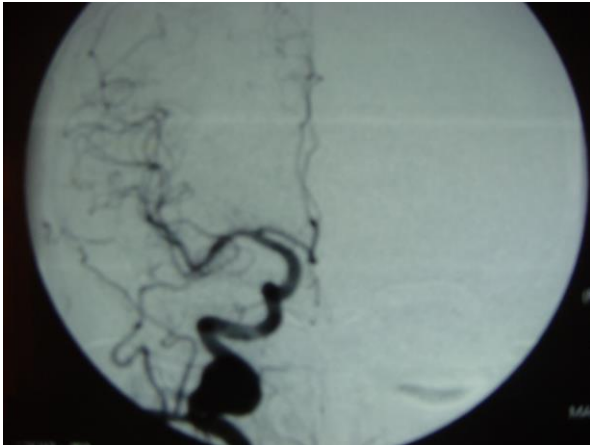


Figure VI. Digital subtraction angiography: Saccular aneurysm on petrosal segment of the right internal carotid artery.

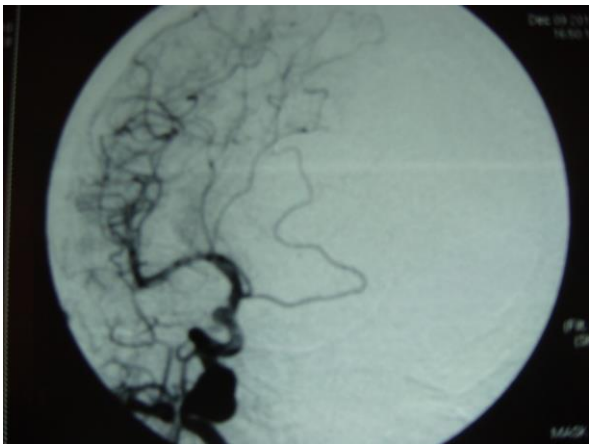


Figure VII. Digital subtraction angiography: Saccular aneurysm on petrosal segment of the right internal carotid artery.

unilateral or bilateral and also accompanied with other neurological findings (1).

Thomson (2) and Lin (3) described an anatomical approach to the differential diagnosis of hypoglossal nerve injury. Supra-nuclear, nuclear and infra-nuclear lesions cause tongue deviation. Nuclear and infra-nuclear lesions will demonstrate lower motor neuron disorder at the same side. If the patients have difficulty in swallowing, the lesion suspected to be peripheral-distal to the point at which C1 fiber joint to hypoglossal nerve. Traumatic skull fracture, vascular etiologies and also granulomatous lesions are also possible causes of this part (4).

There was a few case reports about unilateral HNI. Other etiologies of HNI are; tuberculosis (5), ulcerative colitis (6), occipital condyle fracture (7),

internal carotid artery dissection (8,9), vertebral artery dissection (10), acupuncture (11), mechanical ventilation (4) and other reasons such as; chiari malformation, radiotherapy, post-vaccination, neuropathy, idiopathic (3,12).

Initially, we considered the possibility of chronic arteriolar dissection in the first, and motor neuron disease in the second patients. Both patients have experience of pain on the side of lesion spread to the neck, and also both patients have a history of progressive dysphagia and speech impairment that is why we consider the possibility of tumor, vascular abnormalities on the way of intracranial of extra-cranial portion of the hypoglossal nerve.

To our knowledge, although isolated unilateral HNI is very rare due to ICA dissection (8,9,10), there is no case presentation with isolated unilateral HNI due to ICA aneurysm. On the other hand, chordoma of the occipital condyle is also very rare. Most of time it is presented as occipital condyle syndrome. The other causes of occipital condyle syndrome are; traumatic fracture (7), metastatic lesions, granulomatous lesions (Wegener, tuberculosis etc.) (5,13). Chordomas are slow growing, locally aggressive tumors thought to originate from the embryonic notochord. They can be seen along the entire spine from the cranial base to the sacrum and have high recurrence rates and can also rarely metastasize despite having a benign character. If primary chordomas or its metastases have been detected on cranial base that will be able to lead to cranial nerve involvement. The most common findings or symptoms were headache and/or diplopia (14). In our patient chordoma on cranial base was the main reason of the HNI.

Treatment have been done according to etiology in both of patients; resective operation of the chordoma, and endovascular coil embolization, respectively.

Treatments of two patients are certain. Resective operation for the first and endovascular coil embolization for the second patient have been achieved.

Herein we describe two unusual cases with isolated unilateral HNI, which have been not reported before. These cases highlighted the need for importance of neurologic examination and knowledge of differential diagnosis.

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Ethics

Informed Consent: Informed consent forms were signed by the patients.

Copyright Transfer Form: Copyright Transfer Form was signed by all authors.

Peer-review: Internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices: KA, HB, DI, EKB, KMO, YKB, EA, SZ, Concept: KA, HB, DI, EKB, KMO, YKB, EA, SZ, Design: KA, HB, DI, EKB, KMO, YKB, EA, SZ, Data Collection or Processing: KA, HB, DI, EKB, KMO, YKB, EA, SZ, Analysis or Interpretation: KA, HB, DI, EKB, KMO, YKB, EA, SZ, Literature Search: KA, HB, DI, EKB, KMO, YKB, EA, SZ, Writing: KA, HB, DI, EKB, KMO, YKB, EA, SZ.

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