








Case Report

## A rare neurological presentation of Epstein-Barr virus: Isolated hypoglossal nerve palsy in a child

### *Ebstein-Barr virüsün nadir nörolojik bir tutulumu: Bir çocukta izole hipoglossal sinir felci*

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#### ABSTRACT

The isolated unilateral hypoglossal nerve palsy (HNP) is an unfamiliar clinical sign since it is rarely involved in isolation without the simultaneous involvement of other cranial nerves. It warrants a detailed history and clinical assessment. Neurologic manifestations are infrequent in infectious mononucleosis and especially hypoglossal nerve involvement is uncommon in daily practice. We report a 13-year-old girl who complained of deviation of the tongue to one side, trouble pronouncing linguals and difficulty chewing and swallowing with serologic evidence of active Epstein-Barr virus (EBV) infection. She developed transient isolated unilateral HNP manifests as parapharyngeal inflammation. Rarity and evaluation of isolated HNP pay particular attention in the pediatric field. Although HNP is an unusual neurologic complication of EBV, it should be considered in the differential diagnosis in the early evaluation of children.

**Keywords:** *izole palsi, hipoglossal sinir, epstein-barr virus*

#### ÖZET

İzole tek taraflı hipoglossal sinir paralizisi (HSP), diğer kranial sinirlerin eş zamanlı tutulumları olmadan izole olarak nadiren etkilendiği için alışıldık bir klinik bulgu değildir. Detaylı bir hikaye ve klinik değerlendirme gerektirir. Nörolojik bulgular enfeksiyöz mononükleozda sık değildir ve özellikle hipoglossal sinir tutulumu günlük pratikte çok nadirdir. Biz, Epstein-Barr virus (EBV) enfeksiyonunun serolojik bulguları olan, dilde bir tarafa deviasyon, konuşma, çiğneme ve yutma güçlüğü şikayetleri ile başvuran 13 yaşında bir kız hasta sunduk. Parafaringeal inflamasyonun eşlik ettiği geçici izole tek taraflı HSP ile seyretti. Pediatriye izole HSP nadir görülür ve değerlendirilmesi önem arz eder. Her ne kadar HSP, EBV'nin nadir bir nörolojik komplikasyonu olsa da, çocuklarda ayırıcı tanının değerlendirilmesinde düşünülmelidir.

**Keywords:** *isolated palsy, hypoglossal nerve, epstein-barr virus*

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**INTRODUCTION**

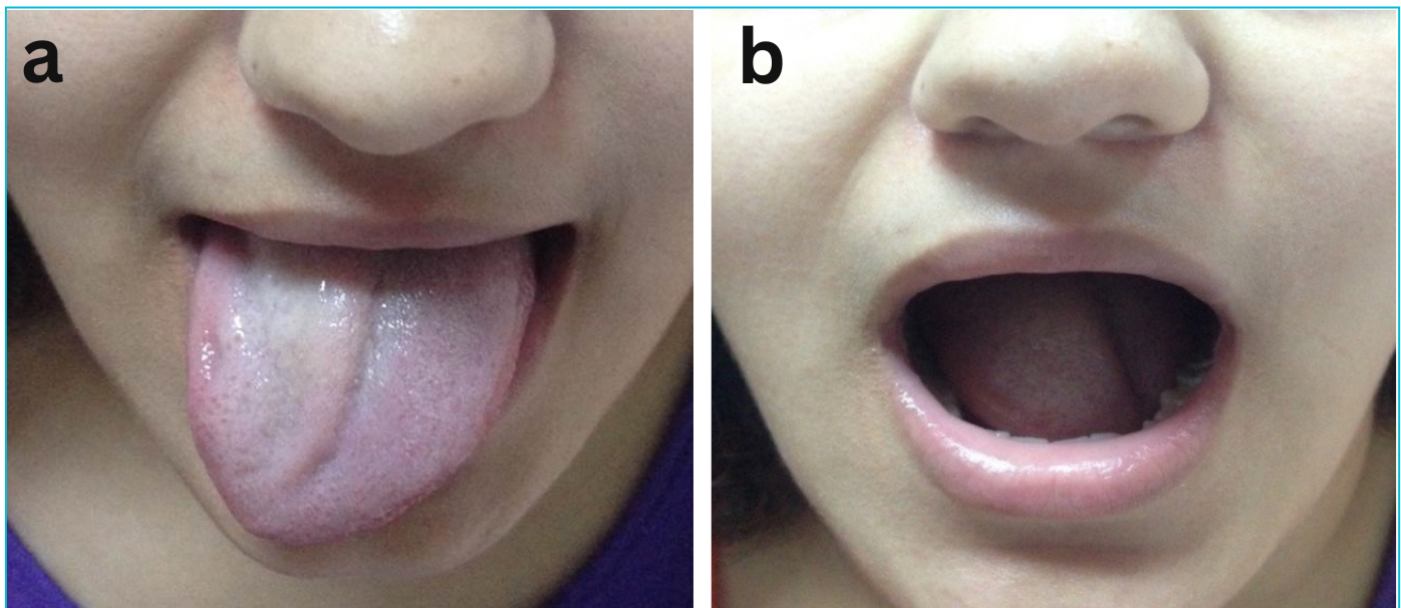
Hypoglossal nerve palsy (HNP) may arise from multiple causes and is frequently associated with other cranial nerve palsies because of its intimate relationship (1,2). Indeed, isolated involvement of the nerve is an exceptional condition with a diagnostic challenge in clinical practice which requires an intensive work-up for a wide variety of causes that may lead to damage of the nerve. Cranial nerve palsies are like an ominous sign because it is indicative of a severe underlying pathology as malignancy, trauma, and stroke. In most adult cases, treatable etiologies are also emphasized, such as infections, autoimmune and vascular pathologies (3,4).

In a few case reports, viral infections, such as Epstein-Barr virus (EBV), varicella-zoster, herpes simplex, measles, and coxsackie A9, have been reported as a responsible etiology for isolated cranial nerve palsy so far (5,6).

We herein reported a 13-year-old girl of isolated, unilateral hypoglossal nerve palsy whose extensive investigations noted serologic evidence of active EBV infection accompanying a parapharyngeal inflammation.

**CASE REPORT**

A 13-year-old girl was admitted to our hospital suffering from difficulty chewing, and swallowing and an uncomfortable sensation on her tongue with a history of slurred speech after five days of ear and neck pain. Her previous medical history was normal, with no recent trauma. Neurological examination revealed that she could not stick out her tongue straight, had slurred speech with the feeling of a thick or clumsy tongue and had difficulty protruding her tongue to the left with muscle bulk loss; these findings were consistent with a unilateral lower motor neuron hypoglossal nerve (Fig 1 a,b). Actually, examination of the other cranial nerves and deep tendon reflexes were normal; the rest of the neurological evaluation were unremarkable. Additionally, her pain and swelling on the right side of her neck lead our attention through a possible inflammation. We noted a temperature of 37.3°C, blood pressure of 110/76 mm/hg and without any meningeal irritation signs, lymphadenopathy, hepatomegaly or splenomegaly.



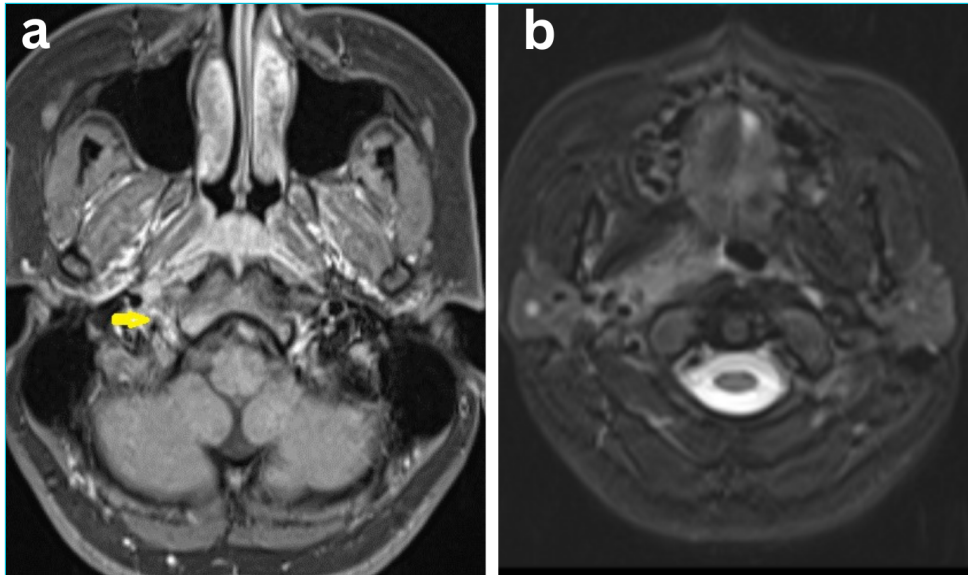
**Figure 1.** a) Right side deviation of the tongue on protrusion, b) Right tongue mounding at rest

Initial investigations comprising a complete blood count, urea, creatinine, uric acid, electrolytes, transaminases, and blood sugar tests were normal. We performed erythrocyte sedimentation rate, C-reactive protein, antinuclear antibody, anti-dsDNA, and vitamin B12, but all these parameters were normal or negative. Viral serology showed us that EBV VCA IgM was positive, and a

positive EBV DNA PCR value of 2510 copy/ml was justified the active EBV infection. Subsequently, with the presentation of isolated HNP and her complaints of right-sided neck pain, a magnetic resonance imaging (MRI) of the head and neck was performed. It revealed an inflammatory involvement of predominantly prestyloid parapharyngeal space. Post-styloid parapharyngeal space also extended with this inflammatory process.

The inflammatory process was infiltrating the hypoglossal nerve at the medial part of the jugular bulb, dorsal to the internal carotid arteries on the right side ipsilateral of the lesion (Fig 2 a,b,c). Parapharyngeal inflammation due to EBV and/or co-infection of parapharyngeal space was diagnosed and a trial of antibiotics

(ceftriaxone 50 mg/kg/d and clindamycin 20 mg/kg/d) was prescribed for ten days. During the follow-up period, the patient's dysarthric speech, and difficulty swallowing and neurological signs of HNP regressed within three weeks.



**Figure 2.** a) Axial contrast-enhanced fat-saturated T1-weighted image and b) Axial fat suppressed T2-weighted image show infiltration of hypoglossal nerve with the parapharyngeal space inflammation.

## DISCUSSION

The hypoglossal nerve provides motor innervation to the tongue playing an essential role in the speech articulation. Unilateral HNP manifests as unilateral atrophy of the tongue, dysarthric speech, and difficulty swallowing as core symptoms and mostly accompanying other cranial nerve involvement because of its intimate relationship (3,4). Since its diverse etiologies, such as neoplasm, trauma, aneurysm, infection and pathologies, are located at multiple sides along its pathway, evaluation of these individuals is a challenge in clinical practice.

In the literature, isolated, unilateral HNP cases are reported in case reports and mostly pronounced as heralds of malignancy in rare case series (4). However, case series of isolated HNP by Combarros et al.(7) reported their nine cases with isolated HNP diagnosed idiopathic of four patients, while the others were with metastatic diseases and brain malformations. Sharma et al.(6) emphasized in their large case series the treatable conditions as tuberculosis and sarcoidosis other than neoplastic pathologies reported in the previous studies as the most common causes. Recently, it is

mostly underlined the need for an extensive diagnostic work-up for potentially treatable etiologies in cases of isolated HNP.

Neurological complications of EBV are a well-known cause of central nervous system infections presenting with a wide range of clinical spectrum; cranial nerve palsies, mono-neuropathies, and many other neurologic, such as acute meningitis and/or encephalitis, acute cerebellar ataxia, and acute disseminated myelitis (8,9). However, isolated cranial nerve palsy accompanying EBV infection has rarely been reported (10). The findings of our neurological examination were unremarkable except for right-sided HNP and associated dysarthria of our patient; therefore, a diagnosis of isolated, unilateral HNP was made. Although the physical examination was not supportive of EBV infection without organomegaly and lymphadenopathy, the serological markers were justified the active EBV infection and neuroimaging showed us a parapharyngeal inflammation as the only cause of this clinical condition. HNP regressed in three-week period after medical treatment. In this case study, we proposed that treatable conditions should be investigated in isolated HNP patients before

extensive diagnostic tests for malignancies in children.

We herein reported a case of an isolated, unilateral HNP in a child both with clinical process and outcome. Clinical and neuroimaging studies led to the diagnosis of active EBV infection accompanying a parapharyngeal inflammation with clinical presentation of isolated HNP. Although isolated HNP caused by EBV and/or parapharyngeal infections is a rare and worrying condition in the clinical ground, early in evaluating children with HNP, serology and neuroimaging should be considered in the differential diagnosis.

#### **Patient Consent Form / Hasta Onam Formu**

The parents' of this patient consent was obtained for this study.

#### **Conflict of Interest / Çıkar Çatışması**

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

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