

Isolated hypoglossal nerve palsy in a child

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We report an 11-year-old boy who had isolated hypoglossal nerve palsy one week after symptoms and signs of urticarial lesions. Neuroradiological examinations and other investigations for etiology of hypoglossal nerve palsy and urticaria were normal. We suggest that all patients with hypoglossal palsy must be carefully evaluated for atypical findings and etiologies.

Key words: hypoglossal nerve palsy, urticaria.

Except for paresis of the facial nerve, mononeuropathies in childhood are very rare. Isolated hypoglossal nerve palsy is a rare condition and may be due to vaccination, aneurysms, trauma, dislocation of vertebrae, intracranial tumor or infectious processes such as infectious mononucleosis. In the literature, there are a few reports about isolated, unilateral hypoglossal nerve palsy¹⁻⁹.

In this report, we present a patient with isolated hypoglossal nerve palsy following urticarial lesions which might have been due to preceding viral infections.

Case Report

An 11-year-old male patient was well until four weeks before admission, when he experienced symptoms and signs of urticaria on his body and in his oral cavity. Corticosteroid and anti-histaminic therapy was given orally in a local hospital. One week following the onset of urticaria, the patient noticed his tongue was atrophic and deviated to the right side on protrusion. There was no history of trauma, vaccination, viral infection, stroke or surgery. Physical examination revealed blood pressure of 110/80 mmHg; skin was normal in appearance. He did not have lymphadenopathy or organomegaly. He was alert and cooperative, and his neurological examination was completely normal except for findings in the tongue. The right side of the tongue was atrophic and deviated to the right side on protrusion (Fig. 1), and fasciculations were noted.



Fig. 1. The right side of the patient's tongue is atrophic and deviates to the right side on protrusion.

On laboratory investigations: the routine complete blood cell count, peripheral blood smear, urinalysis findings, biochemical investigations, slide test for infectious mononucleosis (monospot), herpes simplex virus IgM and G, anti cytomegalovirus (CMV) IgM and G, and Epstein-Barr virus (EBV) VCA IgM were negative. Epstein-Barr virus VCA IgG was positive in titers: 3.1 (N:<1.0). VDRL, ANA, anti dsDNA, ASO, CRP, Latex, Hbs Ag and other hepatitis markers, blood immunoglobulin levels, virus panels including respiratory syncytial virus, adenovirus, influenza A and B, and parainfluenza were normal. Throat swab culture for streptococcal pharyngitis and skin prick tests for common allergens were negative. Clq esterase level was normal. No parasite was detected on stool investigation.

Electrophysiologic evaluation was performed three weeks after the onset of symptoms using Nihon Kohden Neuropach 2 EMG equipment. Compound muscle action potentials (CMAPs) were recorded from lingual muscles with surface clip electrodes. Active recording electrode was placed over ventral surface of tongue (middle of tongue surface, 1 mm from midline) and reference electrode 3 cm proximal. The stimulus was supramaximal, 0.2 ms in duration, and cathode proximal. Nerve conduction studies of left hypoglossal nerve were normal, but right side showed markedly low CMAPs amplitudes with prolonged distal latency. Concentric needle electromyography (EMG) of the right side revealed 2+ fibrillation and positive sharp waves with no recruited motor units. Electrodiagnostic studies repeated five months later showed that right hypoglossal CMAPs amplitudes were mildly increased. On needle EMG rare recruited motor units action potentials appeared with 1+ fibrillation.

The chest roentgenography, X-ray of the skull and sinus roentgenography were normal. Cranial and cervical magnetic resonance imaging (MRI) did not reveal any brain tumor or ischemic lesion. On the MRI, hypoglossal nerve and hypoglossal foramen were normal. He was treated with multivitamins, and examined periodically every other week. Now, at the 20th week of therapy, minimal recovery was recorded clinically and on EMG.

Discussion

Hypoglossal nerve motor composition is highly complex and not fully understood, with the nucleus consisting of four topographically distinct subnuclear columns⁴. Peripheral lesions of the hypoglossal nerve are generally classified into four categories: extramedullary intracervical lesions, hypoglossal foramen lesions, extracranial lesions of the XIIth nerve at the base of the skull, and cervical hypoglossal nerve lesions⁷. In our patient, investigations of the cervical base of the skull showed no abnormal findings.

Isolated, unilateral hypoglossal nerve palsy is rare and usually results from vaccination, carotid and vertebral artery aneurysms, trauma, dislocation of the first cervical vertebra, intracranial neurilemoma or from infectious mononucleosis^{1-3,5,6,8,9}. Keane⁴ described the causes and characteristics of hypoglossal nerve

palsy in a review of 26 years of personal experience. In this patients, the most common cause was tumors and the second most common gunshot wounds. Other causes included stroke, hysteria, multiple sclerosis, surgery, Guillain-Barré neuropathy and infection. Keane could not describe any specific cause in 3% of his patients.

Urticaria affects up to 20% of the population at some time. Acute urticaria may be produced by a number of factors such as infectious agents, foods, insect bites, inhalants, drugs, physical factors, collagen vascular diseases, genetic disorders, neoplasms and hyperthyroidism¹⁰. The most common infectious causes of the urticaria are streptococcal pharyngitis, sinusitis, hepatitis, mononucleosis, viral infections and parasites. In our patient, we could not establish the cause for urticaria except for EBV IgG positivity. Investigations for collagen vascular diseases were normal. There was no history of drugs, insect bites, inhalants, physical agents, nor any notable family history. In our patient, we thought that the urticaria might have been due to preceding viral infections.

In cases of infectious mononucleosis, the incidence of dermatitis is 3% to 19%. It appears during the first few days of illness. Sometimes urticarial or scarlatiniform eruptions are seen¹¹. Neurologic symptoms may occur following acute EBV infection in the absence of symptomatic infectious mononucleosis¹². Neurologic complications of EBV infection include lymphocytic meningitis, encephalomyelitis, polyneuritis and mononeuritis⁶. While any of the cranial nerves may be affected during EBV infection, palsies of cranial nerve VII are most common. Other forms of cranial nerve involvement include anosmia, bilateral sensorineural hearing loss, hypoglossal nerve palsy, and extraocular muscle palsies. In younger children, EBV infections may not have characteristic symptoms and heterophil antibody titers may be negative. The acute illness may be diagnosed if VCA-specific IgM is present in serum. VCA IgM responses disappear after several months, whereas VCA+IgG levels persist for life¹¹. In our patient, VCA IgG level was increased but VCA IgM level was normal. For that reason we cannot say that the urticaria and hypoglossal palsy were due to EBV infection.

Only 15% of the patients with hypoglossal paralysis recovered completely or nearly completely. A rapid onset of hypoglossal nerve

palsy resolves in a minority of patients without specific diagnosis or treatment^{1,7}. In our patient, we noted minimal recovery at the 12th week. On the EMG reevaluation there was recovery at needle EMG findings but there was no change at nerve parameters.

As a result, we believe that all patients with hypoglossal nerve palsy must be carefully evaluated for systemic disorders and infectious agents such as EBV others.

REFERENCES

1. Afifi AK, Rifai ZH, Faris KB. Isolated, reversible, hypoglossal nerve palsy. *Arch Neurol* 1984; 41: 1219-1220.
2. De Simone PA, Synder D. Hypoglossal nerve palsy in infectious mononucleosis. *Neurology* 1978; 28: 844-847.
3. Felix JK, Schwartz RH, Myes GJ. Isolated hypoglossal nerve paralysis following influenza vaccination. *Am J Dis Child* 1976; 130: 82-83.
4. Keane JR. Twelfth-nerve palsy. *Arch Neurol* 1996; 53: 561-566.
5. Parano E, Giuffrida S, Restivo D, et al. Reversible palsy of the hypoglossal nerve complicating infectious mononucleosis in a young child. *Neuroped* 1998; 1: 46-47.
6. Sibert JR. Hypoglossal nerve palsy complicating a case of infectious mononucleosis. *Postgrad Med J* 1972; 48: 691-692.
7. Sugama S, Matsunaga T, Ito F, et al. Transient unilateral, isolated hypoglossal nerve palsy. *Brain Dev* 1992; 14: 122-123.
8. Wright GD, Lee KD. An isolated right hypoglossal nerve palsy in association with infectious mononucleosis. *Postgrad Med J* 1980; 56: 185-186.
9. Giuffrida S, Lo Bartolo ML, Nicoletti A, et al. Isolated unilateral, reversible palsy of the hypoglossal nerve. *Eur J Neurol* 2000; 7: 347-349.
10. Zacharisen MC. Pediatric urticaria and angioedema. *Immunol Allergy Clin North Am* 1999; 19: 363-381.
11. Katz BZ, Miller G. Epstein-Barr virus infections. In: Katz SL, Gershon AA, Hotez DJ (eds). *Krugman's Infectious Diseases of Children* (9th ed). St. Louis: Mosby-Year Book Inc; 1992: 98-115.
12. Ross JP, Chen JI. Epstein-Barr virus. In: Scheld WM, Whitley RJ, Ourach DT (eds). *Infections of the Central Nervous System*. Philadelphia: Lippincott-Raven; 1997: 117-124.