



Ocular, speech, and swallowing problems in a 9-year-old boy: A rare case of polyneuritis cranialis

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Multiple cranial neuropathy is defined as the dysfunction of two or more cranial nerves which can be caused by traumatic, toxic, vascular, neoplastic, infectious, and inflammatory reasons.¹ A common cause of polyneuropathy in pediatrics is neuropathies such as Guillain-Barre syndrome (GBS). GBS is an autoimmune neuropathy triggered by infectious causes such as viral respiratory or gastrointestinal diseases. The incidence of GBS is estimated to be 1-2 in 100000 of the general population. The manifestation of GBS is the progressive ascending weakness of the limbs and the absence of the deep tendon reflexes. During the course of GBS, facial nerve palsy, pain, autonomic dysfunction, paresthesia, numbness, and respiratory failure may occur. The diagnosis is

based on the clinical characteristics, elevated levels of protein in cerebrospinal fluid (CSF) without pleocytosis, and electrophysiologic findings.²

Miller-Fisher syndrome (MFS) is a variant of GBS that involves the cranial nerves and is presented by the triad of areflexia, ophthalmoplegia, and ataxia.³ Cranial nerve involvement is a common finding in GBS but multiple cranial nerve palsy is rare. This type has been mentioned in the literature as polyneuritis cranialis (PNC) and accounts for 3%-5% of the variants. Acute multiple cranial neuropathies without limb involvement are very rare.⁴

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Here, we presented a 9-year-old boy with post-infectious PNC with full recovery after intravenous immunoglobulin (IVIG).

A 9-year-old boy presented to the emergency department of Children’s Medical Center, Tehran, complaining of ptosis and visual, speech, swallowing problems since five days ago. On physical examination, he was unable to move his eyes in all directions. The pupillary reflexes were intact. He was unable to close his eyes completely. He had bilateral weakness of the masseter and temporal muscles, nasal speech, decreased gag reflex, and left deviation of the uvula. His gait, tone, and force of the limbs, deep tendon reflexes, and plantar reflexes were normal. Besides, the sensory exam and bowel and bladder control were intact. The findings were consistent with bilateral symmetrical lower motor neuron facial palsy. He had lower motor neuron type of neuropathy involving the 3rd, 4th, 5th, 6th, 7th, 9th, and 10th cranial nerves. His past medical history was not significant except for a flu-like illness a week and a bloody diarrhea 2 days prior

to the onset of the neurologic symptoms. He mentioned eating a canned tuna fish two weeks ago. Five days before admission, he started to have ocular problems, dysarthria, dysphagia, diplopia, photophobia, and bilateral ptosis.

Electromyography (EMG)-nerve conduction velocity (NCV) reported that compound muscle action potentials (CMAPs) of both facial nerves were low amplitude with normal distal latency; other nerve conduction studies (NCSs) were normal. Needle-EMG of limbs, distal and proximal muscles, and cranial muscles demonstrated a neurogenic pattern only in cranial muscles (tongue, masseter, and facial muscles), with reduced recruitment and spontaneous potentials such as fibrillation and positive sharp waves (PSW).

Brain magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) were normal. All laboratory findings were within normal limits, except for complete blood cells and stool examination, which indicated leukocytosis and white blood cells (WBC) in stool (Table 1).

Table 1. Laboratory findings in patient with polyneuritis cranialis (PNC)

Variable	Result	Normal range
WBC (total)	4-10 × 1000/μl	22.96
Lymphocyte	20-40 × 1000/μl	43.30
Neutrophil	50-70 × 1000/μl	45.70
Hemoglobin	11-16 g/dl	15.70
Platelet	150-450 × 1000/μl	432.00
ESR	0-10 mm/hour	20.00
CRP		Negative
CSF	WBC (mm ³)	1 (lymphocyte)
	RBC (mm ³)	0
	Glucose	68.00
	Protein	40.00
	CSF/culture	Negative
	ACE	23.80
	PANCA	Negative
	CANCA	Negative
	Lupus anticoagulant screen	34.00
	Anti-phospholipid (IgG)	Negative
	Anti-phospholipid (IgM)	Negative
	Anti-B2 glycoprotein (IgG)	Negative
	Anti-B2 glycoprotein (IgM)	Negative
	Anti-cardiolipin (IgG)	Negative
	Anti-cardiolipin (IgM)	Negative
Complement profile	C3	89-195 mg/dl
	C4	10-40 mg/dl
	CH50	70-150 U/ml
	HBsAg	Negative
	HCV Ab	Negative
	HIV Ab	Negative
Stool examination	WBC	< 5/high-power field
	RBC	< 5/high-power field
	Culture	Shigella was detected

ESR: Erythrocyte sediment rate; CRP: C-reactive protein; CSF: Cerebrospinal fluid; ACE: Angiotensin converting enzyme; PANCA: Perinuclear antineutrophil cytoplasmic antibody; CANCA: Cytoplasmic antineutrophil cytoplasmic antibody; HBsAg: Hepatitis B surface antigen; HCV Ab: Hepatitis C virus antibody; HIV Ab: Human immunodeficiency virus antibody; WBC: White blood cells; RBC: Red blood cells; IgG: Immunoglobulin G; IgM: Immunoglobulin M

These findings confirmed an acute denervation process that was compatible with the acute axonal type cranial polyneuropathy. The probable diagnosis based on the provided information was post-infectious cranial polyneuropathy, which was confirmed by electrophysiological evaluation showing acute axonal-type cranial polyneuropathy involving multiple cranial nerves. IVIG with a dose of 2 g/kg for 3 days was administered. After 2 days of starting IVIG, the eye movement and swallowing improved and patient discharged with good condition. After a month, on follow-up visit, all symptoms resolved.

Our patient was a 9-year-old boy presented with ptosis, diplopia, dysphagia, dysarthria, and external eye muscle paralysis. The examination of limbs (force and deep tendon reflexes) was normal and he had intact pupil reflexes and normal visual acuity.

The patient had a history of eating canned food; therefore, botulism was considered among the differential diagnoses. The foodborne type of botulism is the commonest. It can be confused with more common diseases such as myasthenia gravis (MG) and GBS (MFS variant). The symptoms appear within 12 hours to 15 days after ingestion of the contaminated food. Autonomic symptoms such as dry mouth and throat can be the earliest and may be confused with pharyngitis. Clinical signs always start with the involvement of 9th, 10th, 11th, and 12th cranial nerves. Then, the characteristics of a typical disease include external ophthalmoplegia, mydriasis (most of the time fixed), blurred vision, diplopia, and bilateral ptosis. Dysphagia, dysarthria, and dry or sore mouth and throat are very specific neurologic symptoms. In contrast to GBS and MFS (a variant of GBS), botulism does not proceed with a history of infectious illness (e.g., flu-like illness or diarrhea). Moreover, in patients with GBS, the pupil reflexes are intact.⁵ Our patient's pupillary size and reflexes and deep tendon reflexes were intact; therefore, the diagnosis of botulism was not a high probability.

Electrophysiology studies in GBS may show demyelinating and/or axonal involvement. Cranial nerves can get involved in 45%-75% of the patients. The 3rd, 4th, 6th, and 7th cranial nerves (facial and extraocular nerves) are the most

affected and the cranial nerves located at the lower part are the least involved.⁶ In our patient, EMG and NCV studies revealed acute axonal type cranial polyneuropathy.

Cranial nerve involvement is a common finding in GBS but multiple cranial nerve palsy is rare. This type has been mentioned in the literature as PNC and accounts for 3%-5% of the variants. Acute multiple cranial neuropathies without limb involvement are very rare.⁴

PNC can be considered as an oculopharyngeal subtype of GBS, which manifests with ocular and pharyngeal involvement without limb weakness. The 7th, 9th, and 10th cranial nerves are most frequently involved. The flu-like illness and bloody diarrhea in our patient may be coincident findings or be relative to PNC.

Wakerley and Yuki reviewed the previous cases of PNC and found that the 1st and 2nd cranial nerves involvement was not reported and albuminocytological dissociation was noted in 67% of the patients. Etiologies can be attributed to the inflammatory, infectious, or carcinomatous processes involving the nerves (e.g., sarcoidosis), involvement of the meninges (e.g., carcinomatous meningitis), and involvement of the nerves at the skull base (e.g., metastasis). Deep tendon reflexes may be present.⁷

We reported a patient with multiple cranial neuropathy with excellent response to IVIG. PNC should be considered in any patient with multiple cranial nerve involvement. Although it is usually self-limited, early diagnosis and intervention may accelerate the healing process.

Written informed consent was acquired from the patient's family. Research Ethics Committees of Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran, approved the use of humans for this study and the related ID is IR.TUMS.CHMC.REC.1403.017. This study conforms to the requirements of the Declaration of Helsinki (1989).

Conflict of Interests

The authors declare no conflict of interest in this study.

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None.

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