

AN ISOLATED HYPOGLOSSAL NERVE PALSY FOLLOWING PHARYNGITIS: A CASE REPORT

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Background

The hypoglossal nerve innervates the intrinsic and extrinsic muscles of the tongue with the exception of the palatoglossus, which is innervated by the vagus nerve. It has five different segments (medullary, cisternal, skull base, carotid space, and sublingual) and it has a complex course, close to other neurological structures and vessels, that's why hypoglossal nerve palsy (HNP) usually develops in association with other neurological signs and symptoms.

Amro et al. in December 2016 published a retrospective analysis of cases of HNP that had been evaluated at the Mayo Clinic over a 30-year period, a total of 245 patients (54.7% males). In the 49% of cases the HNP presented isolated. Etiologic categories were postoperative (29.3%), idiopathic (15.1%), primary neoplastic (14.2%), metastatic (13.0%), inflammatory (7.3%), radiation-induced (6.1%), traumatic (4.1%), vascular (3.3%), congenital (2.8%), cystic (2.4%), and motor neuron disease-related (1.6%).

Idiopathic IHNP is a rarer finding. It has usually a benign course and the majority of patients have an excellent outcome. Some cases of persistent idiopathic IHNP has also been described in literature.

Case report

A 51 year-old man with isolated XII left nerve palsy presented to the emergency room for acute pharyngitis with plaques associated to neck rigidity. Past medical history was significant only for essential tremor and tonsillectomy. The brain CT scan was normal. Neurological exam was negative except for a tongue left hemiatrophy associated to a normal tongue motility.

We visited the patient for the first time after eight months and we reported a left tongue hemiatrophy associated to fasciculations, an ipsilateral deviation of tongue on protrusion, a contralateral deviation at rest. No other clinical signs were observed.

The patient underwent a range of **haematological investigations** including inflammatory markers, serology for treponema, HIV, HCV, HBV, autoimmunity test and research of tumor markers, all of which were unremarkable.

Otorhinolaryngological evaluation revealed an isolated hypotrophy of the left side of tongue; **fibroendoscopy evaluation** of cordal motility was normal.

MRI of the brain associated to intracranial angiography didn't show parenchymal or vascular abnormalities.

A **chest CT scan** excluded malignancies, tuberculosis or sarcoidosis

Electromyography (EMG) showed signs of chronic denervation associated to collateral reinnervation of left genioglossus associated to a mild diffuse polyneuropathy.

There was no family history of idiopathic IHNP or any other neurological disorders.

After a 18 month follow-up period, the patient's neurological examination remained unchanged.



Possible causes of hypoglossal nerve palsy

POSTOPERATIVE	endarterectomy, glomus jugulare resection
VASCULAR	carotid artery dissection/aneurysm, basilar ectasia, vertebral dissection, dural arteriovenous fistula of the transverse sinus, thrombosis of median branches of vertebral artery, jugular thrombophlebitis, infarct/haemorrhage over the hypoglossal nerve nucleus, cavernoma
TRAUMATIC	head and neck trauma with or without fracture
NEOPLASTIC	metastatic disease at base of skull, meningioma, glomus tumor, medullary segment glioma, nasopharyngeal carcinoma, nerve sheath tumor, sublingual segment carcinoma, lymphoma
INFLAMMATORY	sarcoidosis, vasculitis, multiple sclerosis, rheumatoid arthritis, Guillan-Barré neuropathy, Wegener's granulomatosis
INFECTIOUS	post-retropharyngeal infection, tuberculosis, meningococcal meningitis, dental infection, abscess, mononucleosis
CONGENITAL	Arnold-Chiari malformation
OTHERS	motor neuron disease radiation-induced syringobulbia, Diabetes mellitus, hysteria, branchial cyst
IDIOPATHIC	No cause found

Conclusions

The temporal association suggests a casual relationship between the pharyngitis and the hypoglossal palsy.

To our knowledge, only two cases of post viral HNP are reported in literature (Naik et al, Carra-Dallière et al). We found more HNP linked to bacterial infections (tuberculosis, meningococcal meningitis, dental infection). In most cases patients showed a spontaneous recovery.

Idiopathic HNP should remain a diagnosis of exclusion after a complete clinical, laboratory and radiological investigation.