Idiopathic Horner’s Syndrome: An Enigma

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ABSTRACT

Horner’s syndrome (Bernard-Horner’s syndrome or oculosympathetic palsy) is a clinical syndrome caused by paralysis of the cervical sympathetic trunk. It is characterized by miosis, ptosis, enophthalmos and anhidrosis. Most of the cases are due to vascular causes, trauma or cancer. Occasionally, Horner’s syndrome is encountered in a patient where no cause can be ascertained. Here, we describe one such case.

Keywords: Enophthalmos, oculosympathetic palsy, miosis, ptosis

Horner’s syndrome (Bernard-Horner’s syndrome or oculosympathetic palsy) is a clinical syndrome caused by paralysis of the cervical sympathetic trunk. It is characterized by miosis, partial ptosis of the eyelid, enophthalmos and sometimes hemifacial anhidrosis. Most of the cases are due to brainstem strokes, trauma, apical lung tumors or vascular malformations. Although a hereditary autosomal dominant form is known, idiopathic Horner’s syndrome is only seldom encountered. Here, we describe this presentation in a 36-year-old female.

CASE REPORT

The patient presented to the Medical OPD with complaints of left-sided facial dryness and drooping of left eyelid more noticeable in the recent few months. On examination, typical features of Horner’s syndrome were clearly appreciable on the left side (Figs. 1 and 2). Rest of the general as well as systemic physical examination was unremarkable. A relevant history directed to all possible causes was ardently sought but was noncontributory. A detailed lab profile followed by a CXR-PA, ECG, CECT of the chest and a CEMRI with MRA (intracranial vessels) of the brain were done, all yielding normal results. We resorted to pharmacological testing of the involved pupil with 2% cocaine to confirm our observation, and a 1% hydroxyamphetamine test localized the lesion to the preganglionic pathway. A diagnosis of idiopathic Horner’s syndrome was thus entertained.

DISCUSSION

Horner’s syndrome is usually acquired as a result of a pathology but may also be congenital, iatrogenic and very rarely idiopathic. Some of the common causes and associations are elaborated below:

- Lateral brainstem strokes
- Cluster headache - combination termed Horton’s headache
- Trauma-base of neck, usually blunt trauma, sometimes surgery
- Middle ear infection
- Tumors - often bronchogenic carcinoma of the superior fissure (Pancoast tumor) on apex of lung

Figure 1. Left hemifacial anhidrosis, ptosis and enophthalmos.

Figure 2. Anisocoria with a constricted left pupil.
Thoracic aortic aneurysm
Neurofibromatosis type 1
Goiter
Dissecting aortic aneurysm
Thyroid carcinoma
Multiple sclerosis
Carotid artery dissection
Klumpke paralysis

Keane has provided data on the relative frequency of the lesions causing Horner’s syndrome. In 100 successive cases, 63 were of central type due to brainstem strokes, 21 were preganglionic from trauma or tumors of the neck, 13 were postganglionic due to miscellaneous causes, and in three cases the localization could not be determined. Signs found in all patients on affected side of face include ptosis (drooping upper eyelid from loss of sympathetic innervation to the Müller or superior tarsal muscle), upside-down ptosis (slight elevation of the lower lid), miosis and dilation lag. Enophthalmos (the impression that the eye is sunk in) and anhidrosis on the affected side of the face, loss of ciliospinal reflex and blood shot conjunctiva may occur depending on the site of lesion. Also flushing of the face is common on the affected side of the face due to dilation of blood vessels under the skin. The presence, absence and/or location of anhidrosis is an important localizing sign that may be elicited from the history, thus anhidrosis is generally not found in lesions distal to the carotid bifurcation.

Although Horner syndrome is commonly an incidental finding related to a benign cause, it occasionally may be a manifestation of a serious and life-threatening disorder. Careful evaluation to rule out such life-threatening disorders is therefore of utmost importance.

Lab studies in general do not play a role in the diagnosis and management of Horner’s syndrome. However, depending on the localization and suspected etiology, lab tests which one may consider, in conjunction with appropriate medical consultation, include the following: CBC count, fluorescent treponemal antibody absorption (FTA-ABS) test, Veneral Disease Research Laboratory (VDRL) test, purified protein derivative (PPD) placement and/or urine test (i.e., vanillylmandelic acid [VMA], homovanillic acid [HVA]) to rule out neuroblastoma in pediatric Horner’s syndrome.

Imaging studies may be ordered in conjunction with appropriate medical and/or surgical consultation depending on the localization and suspected etiology. Such studies may include MRI/MRA, angiography, extracranial Doppler ultrasound and/or chest X-ray.

Pharmacologic testing is very helpful in the diagnosis of Horner’s syndrome and localization of lesions causing Horner’s syndrome. Cocaine inhibits the re-uptake of norepinephrine from the synaptic cleft. Two drops of 4% or 10% cocaine solution are instilled into each eye.

Cocaine instilled in an eye with intact sympathetic innervation causes the pupil to dilate. A sympathetically denervated pupil dilates poorly to cocaine, regardless of the level of the sympathetic interruption because of the absence of endogenous norepinephrine in the synapse. A dilatation response to 1% hydroxyamphetamine 24 hours after the cocaine test localizes the lesion to the preganglionic system.

No definitive treatment is possible in idiopathic Horner’s syndrome. If cause is found out treatment is aimed at the same. Neurologic or neuro-ophthalmic consultation may be considered as a serious underlying cause may sometimes be overlooked.

REFERENCES