Abstract

Horner’s syndrome is an oculosympathetic paresis resulting from damage to the sympathetic innervations pathway to the eye and adnexa. This paper discusses the diagnosis of a Horner’s syndrome occurring after an epidural procedure.

I. Case History

Patient Demographics: 36 year old Caucasian female

Chief Complaint: Sudden onset of “droopy” right eye immediately following the birth of her daughter by caesarian section, and pupils that are two different sizes.

Ocular History: Unremarkable except for successful LASIK surgery on both eyes in 2006

Medical History and Medications:

Patient is negative for systemic hypertension, diabetes, thyroid disease, AIDS, HIV, and Hepatitis C, or multiple sclerosis. She is positive for droopy right eyelid, anisocoria, blood in her urine immediately after the birth, mild anemia during this pregnancy and a hairline fracture in her left arm within the past twelve months. She reported no hospitalizations, with the exception of this birth and a previous cesarean section in 1996. Patient denies any neck manipulation, headaches, migraines, double vision, eye pain, or trauma. She has no hoarseness, trouble swallowing, trouble breathing, fatigue, or tingling/numbness of the face or extremities. She notes a temperature difference on the right side of her face. She denies any medications except for the epidural prior to the C-section and vicodin given after the birth for pain and she is not breastfeeding. She is positive for allergy to penicillin and Sulfa drugs.

II. Pertinent Findings

Visual acuities were 20/20 in both eyes, uncorrected. Color vision and Amsler grid were all normal. Margin to reflex distance, without brow fixation was 2.0 mm in the right eye, and 2.5 mm in the left eye. The patient had a high lid fold on the upper right eyelid. Pupils were round in the light. The right eye was measured at 3.0 mm and the left was 4 mm. In the dark, the right pupil was 4mm, and the left was 5.5mm, with a dilation lag noted in the right eye (not quantified). Direct and consensual responses were +2 in both eyes, with no afferent pupillary defect. Ocular motilities were full and accurate with no nystagmus. She had a small 2 pd esophoria at distance and a small 2 pd exophoria at near. NPC was 10 cm. No cranial nerve paresis was found.

Slit lamp examination was unremarkable for both eyes, with no heterochromia. Intraocular pressures were 15 mmHg for the right eye and 16mmHg for the left eye at 9:56 AM. A dilated fundus exam revealed crowded, elevated, pink optic nerve heads with cup to disc ratios of .1/.1 in each eye. A spontaneous venous pulse was noted in each eye. The maculae were unremarkable in each eye.
Retinal examination revealed no holes, no tears, and no retinopathy. A Humphrey 30-2 size III SITA fast visual field was performed and was unremarkable.

A 1% hydroxyamphetamine bromide (Paredrine, one drop in each eye) test was negative, indicating a central or preganglionic lesion. MRI and MRA results were both normal. At all subsequent exams, the patient’s condition has remained essentially unchanged.

III. Differential Diagnosis

1. Physiological Anisocoria
2. Horner’s Syndrome

IV. Diagnosis and Discussion

Patient was diagnosed with Horner’s Syndrome related to the epidural she received prior to her C-section.

Horner’s syndrome, or oculosympathetic palsy, is a neurologic syndrome resulting from damage to the sympathetic nerves supplying the eye. Causes range from benign to life-threatening, requiring a systematic approach to diagnostic evaluation. The interruption of the sympathetic pathway can occur at any point along the complex path from the brain to the eye. The first-order neuron begins in the hypothalamus of the brain and runs down the spinal cord to the ciliospinal center of Budge, at approximately the level of C8 to T2, and on occasion, as low as T4. The second-order neuron travels from the sympathetic trunk, through the brachial plexus, over the lung apex, synapsing in the superior cervical ganglion near the bifurcation of the common carotid artery. The third-order neuron leaves the superior cervical ganglion to form a plexus surrounding the internal carotid artery, ascends and travels through the cavernous sinus in close relation to cranial nerve six. The pathway then joins the ophthalmic division of cranial nerve five (trigeminal nerve). The sympathetic branches follow first the abducens nerve and then the first branch of the trigeminal nerve before reaching the pupil dilator muscle and the Muller’s muscle.

The classic signs of a Horner’s syndrome are miosis, ptosis, and ipsilateral anhidrosis. The degree of anisocoria is greater in the dark than in the light. There is an associated dilation lag. The affected pupil dilates more slowly, (by 15-20 seconds) than the normal pupil. Because of this, the anisocoria is greater at 5 seconds in the dark (time for the normal pupil to dilate) than at 20 seconds.

The ptosis is minor (around 2 mm or less) because it results from paralysis of the Muller’s muscle, a minor player in lid elevation. There is often an accompanying “upside down ptosis”, wherein the lower lid is elevated slightly due to denervation of the small smooth muscles of the lower lid. This narrows the palpebral fissure and creates an apparent enophthalmus.

Anhidrosis may be present in central or preganglionic lesions. The sympathetic fibers responsible for facial sweating and vasodilation branch off at the superior cervical ganglion from the remainder of the pathway. Therefore, the anhidrosis is not present in postganglionic lesions. Acute features of sympathetic disruption can also include ipsilateral conjunctival injection, nasal stuffiness, and increased near point of accommodation.
Central and preganglionic lesions tend to be more serious. They may be caused by cerebrovascular
infarcts, basal skull tumors, pituitary tumors, basal meningitis, multiple sclerosis, neck trauma,
pancoast tumor, aortic aneurism, neuroblastoma, or surgical injury, to name a few.6 Central Horner’s
syndrome usually presents with accompanying neurological findings, such as diplopia due to cranial
erve paresis found with vertebrobasilar dissection.

Second order neuron lesions, often called “Silent Horner’s”, represent the majority of preganglionic
cases. They are often due to lung disease in adults and neuroblastoma in children. Occasionally,
preganglionic Horner’s syndrome can accompany the administration of spinal or epidural anesthesia
during child birth.1,2,3,4,6,8,9 While the exact mechanism is not known, it is believed to be due to a high
rostral spread of local anesthetic acting on the preganglionic neurons as they exit the spinal cord to
the sympathetic chain. It has been posited that small amounts of the anesthesia may be driven
rostrally by intense labor and by Valsalva maneuver in the second stage of labor.8

Postganglionic lesions are usually painful and due to carotid dissection, cluster headache or neck
trauma. Carotid artery dissection is by far the most common.6

Diagnosis and localization are dependent on clinical features, pupillary function, pharmacologic
testing, and imaging. Because patients are often asymptomatic, the initial step is to differentiate
between simple physiological anisocoria and Horner’s syndrome. Often this can be done just by
observing the dilation lag.7 In cases where the results are inconclusive, the common next step is
cocaine testing. A drop of cocaine (4-10%) is placed in each eye, and the anisocoria is measured 50-
60 minutes later.7 Cocaine blocks the reuptake of norepinephrine released by the nerve ending,
resulting in dilation of a normal pupil. In the case of oculosympathetic denervation, no
norepinephrine is released, so blocking its reuptake would have no effect.10,11,12 The Horner’s pupil
would not dilate.

In recent years, the use of alpha-agonist apraclonidine (Iopidine) has increased as an alternative to
cocaine due in part to the difficulties surrounding cocaine procurement and storage.10,12,13,13 Working
on the theory that the pupillary dilator muscle and Muller’s muscle in a Horner’s syndrome have a
hypersensitivity of α1 receptors, apraclonidine will dilate a Horner’s pupil and temporarily reduce the
ptosis, but have no effect on the normal pupil.10,12,13,14,15

Once diagnosed, the Horner’s syndrome lesion can be localized with 1% hydroxyamphetamine
hydrobromide (Paredrine). Paredrine releases norepinephrine from the nerve endings, causing
dilation in normal eyes. In a postganglionic lesion, the nerve endings themselves are destroyed, so
there is no norepinephrine to be released, and no dilation occurs.16

Despite having localized the lesion pharmacologically, imaging is usually necessary. Central and
preganglionic lesions require brain and spine MRI and an MRA. Preganglionic lesions may require a
chest CT or X-Ray.6,16 In postganglionic lesions, an MRA can identify 94% of carotid dissections,
allowing intervention and possible avoidance of ischemic damage from thromboembolism.6,16 A
brain MRI may also be necessary to look for a cavernous sinus pathology.
V. Treatment and Management

Horner’s syndrome itself is not a treatable condition, but rather an indication of an underlying problem. Treatment is therefore aimed at the pathology. The ptosis may be treated surgically if it is sufficiently bothersome.

As Horner’s syndrome can signal a vast range of pathologies, some life threatening, its timely diagnosis and localization of the lesion are of great importance.

VI. Conclusion

In the case of our patient, as in many cases, the exact etiology remains unknown, and becomes one of exclusion. There is no room for assumption. Although Horner’s syndrome can be a complication of spinal or epidural anesthesia, it should never be assumed to be so. Each case should be thoroughly investigated to rule out any co-morbidity.

References

