

A case of postganglionic Horner syndrome after catheterization of internal jugular vein confirmed with pharmacological tests

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Horner syndrome, consisting of ipsilateral miosis, upper eye lid ptosis, and facial anhidrosis, has been reported as a rare complication of internal jugular vein catheterization.

In this paper, we describe a nine-year-old girl presenting with postganglionic Horner syndrome, in whom right-sided ptosis and miosis occurred after ipsilateral percutaneous internal jugular venous catheterization.

Key words: catheterization of internal jugular vein, Horner syndrome.

Horner syndrome, characterized by the classic triad of ipsilateral miosis, mild upper eyelid ptosis and facial anhidrosis, is caused by interruption of the oculosympathetic pathway at any point between its origin in the hypothalamus and the eye. In children, the etiologies of Horner syndrome have traditionally been subdivided into acquired and congenital causes. The acquired causes consist of sequela of head, neck, and chest surgery, and any neoplasm or infection affecting the oculosympathetic pathway. Meanwhile, birth trauma, neoplasm, and carotid abnormalities are the most commonly identifiable congenital causes¹.

Horner syndrome presents a challenge to the clinician because the causative lesion may involve a first, second, or third-order neuron. Central disorders of the nervous system such as vascular occlusion, particularly in the lateral medulla, tumors, cervical disc disease, and other disorders involving the upper cervical spinal cord are the most common causes of first-order neuron lesions. Second-order neuron lesions are caused by apical lung tumors (i.e., Pancoast syndrome), metastases, chest surgery², thoracic aortic aneurysms, or trauma to the brachial plexus. Third-order neuron lesions are mostly due to degenerative changes in the wall of the carotid artery or they develop following vasospasm. Other causes include surgery of the

carotid artery or adjacent structures, internal carotid artery dissection, and extension of tumors such as nasopharyngeal carcinoma into the cavernous sinus¹.

Horner syndrome has been reported as a rare complication after internal jugular vein catheterization³⁻⁵. Because of the proximity of arterial and cervical sympathetic nerve structures to the internal jugular vein, local complications during insertion can lead to ipsilateral injury. Here, we report a case of postganglionic Horner syndrome, confirmed by pharmacological localization, developing after otherwise uneventful percutaneous insertion of a catheter via the ipsilateral internal jugular vein in atrial septal defect surgery.

Case Report

A nine-year-old girl with high venous atrial septal defect, who underwent atrial septal defect closure surgery, was consulted to our clinic because of the ptosis noticed on the first postoperative day. Ophthalmological examination revealed 2 mm ptosis of the right upper lid and accompanying miosis (Fig. 1). In bright illumination, the right pupil was 3.68 mm and the left pupil 4.28 mm in size. In dim illumination, the right pupil measured 3.65 mm and the left 5.61 mm. We used Adobe Photoshop CS (Adobe Systems Inc, San Jose,



Fig. 1. Ptosis of the right upper lid and accompanying miosis.

California, USA) software to measure the pupil diameters⁶. Pupillary light reflexes were normal on both eyes. No masses or hematomas were felt in the right neck.

A drop of 10% cocaine^{4,7,8} was instilled into each eye, and this was repeated five minutes later. After 30 minutes, the left pupil dilated to 6.11 mm while the right pupil remained at 4.26 mm, confirming the presence of a right-sided Horner syndrome (Fig. 2).

The patient was also tested with phenylephrine 1%⁹ and apraclonidine 0.5%^{6,10,11} on separate days, at least two days apart. After pharmacological testing with phenylephrine 1%, the increase in pupil size of our patient was 2.57 mm in the right pupil and 0.93 mm in the contralateral normal pupil, thus indicating a postganglionic lesion (Fig. 3).



Fig. 2. Cocaine test: After 30 minutes, the left pupil dilated to 6.11 mm while the right pupil remained at 4.26 mm, confirming the presence of a right-sided Horner syndrome.



Fig. 3. Phenylephrine 1% test: After the instillation, the increase in pupil size of our patient was 2.57 mm in the right pupil and 0.93 mm in the contralateral normal pupil, thus indicating a right-sided postganglionic lesion.

Our patient showed reversal of anisocoria after the instillation of apraclonidine 0.5%.

Ophthalmological and neurological examinations were otherwise unremarkable. Doppler ultrasound of carotid arteries was normal. CT scans of the chest and neck were normal. The Horner syndrome has persisted during follow-up for 12 weeks.

Discussion

Sympathetic innervation to the eye consists of a three-neuron arc. The first neuron originates in the dorsolateral hypothalamus, descends through the reticular formation of the brainstem and travels to the eighth cervical and fourth thoracic vertebrae of the spinal cord. There, it synapses with second-order neurons, the preganglionic cell bodies of which give rise to axons. These axons pass over the apex of the lung and synapse in the superior cervical ganglion of the sympathetic chain in the neck, where cell bodies of third-order neurons give rise to postganglionic axons that course to the eye through the cavernous sinus. These sympathetic nerve fibers course anteriorly through the uveal tract and join the fibers of long posterior ciliary nerves to innervate the dilator muscle of the iris. Postganglionic sympathetic fibers also innervate the muscle of Müller within the eyelid, which is responsible for initiation of eyelid retraction during eyelid opening. Postganglionic sympathetic fibers, which are responsible for facial sweating, follow the external carotid artery to the sweat glands of the face. Interruption anywhere along this pathway - preganglionic (first or second neurons before the synapse in the superior cervical ganglion) or postganglionic (after exiting the superior cervical ganglion) - will induce an ipsilateral Horner syndrome. Fibers from these axons form the long and short posterior ciliary nerves of the eye^{1,12}.

In an analysis of 216 cases reported by Giles and Henderson¹³, the most common site of involvement was the cervical sympathetic trunk including the postganglionic fibers proximal to their intracerebral course. In this location, neoplasia was the predominant etiologic factor (66.5%), followed by surgical procedures (30%) and trauma (2%). In the literature, Horner syndrome has been rarely reported after internal jugular vein catheterization³⁻⁵.

We aimed to contribute to the literature with the presentation of this case. In our patient, the acute onset of right-sided Horner syndrome after catheterization of ipsilateral internal jugular vein, in the absence of other neurologic findings or mass lesions in the area of the neck and pulmonary apex, strongly implicated that internal jugular vein catheterization was the etiologic factor.

Vaswani et al.³ and Teich et al.⁴ suggested that the potential causes of Horner syndrome during catheterization of the internal jugular vein were trauma or vascular changes affecting either preganglionic or postganglionic fibers. They proposed that the cause might have been direct trauma from the needle or possibly a toxic effect from extravasated fluid or transient occult hematoma within the carotid sheath compromising the vascular supply of the superior cervical ganglion. Boyd et al.¹⁴ reported that multiple needle passes, inadvertent arterial punctures, and overt cervical hematomas together complicate approximately 9.4% of catheterizations.

In our patient, the diagnosis of postganglionic Horner syndrome was confirmed by increased anisocoria in dim illumination, failure of the affected pupil to respond to cocaine 10%, reversal of anisocoria with apraclonidine 0.5%, and denervation supersensitivity of the affected pupil to phenylephrine 1%^{4,6-11}.

Topical cocaine test has been the gold standard for years. A cocaine test that induces 1 mm or more of anisocoria is strongly supportive (>95% probability) of the diagnosis of Horner syndrome. The cocaine test corroborates the diagnosis of Horner syndrome, but does not allow distinction between a central (brainstem or cervical cord), preganglionic (chest or neck), or postganglionic (above superior cervical ganglion) cause of sympathetic denervation.

Because cocaine is a controlled substance, it is difficult to obtain, and because of its short shelf life, a fresh solution must be prepared for each individual patient. Instead of cocaine, apraclonidine 1% or 0.5%, the effect of which is due to denervation hypersensitivity of the postsynaptic α_1 receptor in the pupil dilator muscle, may be used in the diagnosis of Horner syndrome^{6,10,11}.

After the confirmation of the diagnosis of Horner syndrome by either cocaine or apraclonidine test, the second step in the pharmacological approach

is the localization of the lesion. In this step, hydroxyamphetamine, hydroxyamphetamine hydrobromide, and phenylephrine are the diagnostic tests reported to be effective^{15,16}.

Since it has been reported that phenylephrine 1% correlates well with the results of hydroxyamphetamine 1% in localizing the lesion to the postganglionic neuron and is a reliable alternative to hydroxyamphetamine 1%, which is not readily available, we preferred phenylephrine 1%^{9,17}. It is important to remember that phenylephrine 1% and apraclonidine 0.5% can be easily prepared by dilution of stronger concentrations, which are always available in ophthalmologists' offices. With these tests, it is possible to confirm the diagnosis of Horner syndrome and demonstrate a postganglionic lesion.

In patients with an acute onset of ptosis and miosis diagnosed as Horner syndrome after surgery, jugular vein catheterization should be considered as an etiologic factor and pharmacological confirmation can be done by simple tests.

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