Answer to Ophthaproblem *continued from page 439*

1. Horner syndrome

Horner syndrome (HS) is a clinical syndrome associated with damage to the oculosympathetic pathway (OSP), a chain of neurons that supplies sympathetic innervation to ipsilateral ocular and orbital structures including the sweat glands, the pupil dilator muscle, and the retractor muscles of the upper and lower eyelids (superior tarsal muscles).[1] Classically, it produces the triad of mild ptosis, miosis, and anhidrosis of the head and neck.[2] It might also cause iris heterochromia or asymmetric facial flushing if congenital and conjunctival hyperemia if acute.[3,4]

There are many causes of HS, which are categorized according to the location of the defect within the 3-neuron chain of the OSP. The OSP originates in the hypothalamus, where first-order neurons descend through the brainstem to synapse with second-order neurons in the spinal cord at the C8-T2 level.[5] Second-order neurons then ascend in the sympathetic chain through the inferior and middle cervical ganglion to synapse with third-order neurons in the superior cervical ganglion. Finally, the third-order neurons innervate the sweat glands, pupil dilators, and superior tarsal muscles of the upper and lower eyelids.[1]

Common causes of first-order neuron HS include hypothalamic lesions, lateral medullary plate syndrome (Wallenberg syndrome), syringohydromyelia, multiple sclerosis, and spinal cord neoplasms.[1] In addition to miosis and ptosis, first-order neuron lesions might cause ipsilateral anhidrosis of the entire body and commonly present with cerebellar, brainstem, or spinal cord symptoms.[6] Causes of second-order neuron HS include Pancoast tumours (bronchogenic carcinomas of the lung apex), sympathetic schwannomas, neuroblastic tumours, and goiter; second-order neuron lesions typically produce the classic triad of ptosis, miosis, and ipsilateral anhidrosis of the face and neck.[1] Third-order neuron lesions cause ipsilateral anhidrosis of the entire face and neck only if the superior cervical ganglia is involved; otherwise, anhidrosis is observed on the ipsilateral nose and forehead only.[1] Common causes of third-order neuron HS include fibromuscular dysplasia, carotid dissection, and lesions of the skull base, parasellar space, and orbit.[1]

Management

Careful clinical examination can help differentiate among different causes of anisocoria. The physician should examine pupil size in both light and dark conditions. Anisocoria greatest in the light suggests a parasympathetic lesion (such as third cranial nerve palsy), whereas anisocoria greatest in the dark points toward physiologic anisocoria or a sympathetic lesion, such as in the case of HS. Horner syndrome can be differentiated from physiologic anisocoria by the presence of a dilation lag in the dark; that is, the affected eye dilates slower than the fellow eye in suddenly dark surroundings.[1]

Pharmacologic testing can be subsequently performed to confirm the diagnosis of HS and to localize the lesion. The cocaine test involves administering a 4% to 10% cocaine solution into both eyes and checking for pupil dilation.[4] Cocaine blocks the reuptake of norepinephrine into the postganglionic neurons at the synaptic junction; with an intact sympathetic pathway, this results in dilation of the pupil.[4] Little or no dilatation in response to cocaine therefore supports a diagnosis of HS. To differentiate a preganglionic (in reference to the superior cervical ganglia) from a postganglionic lesion, a hydroxyamphetamine test can be performed as well (but not on the same day as the cocaine test).[4] Hydroxyamphetamine releases norepinephrine into the synaptic cleft; if no dilation is seen, the lesion is likely postganglionic (ie, a third-order neuron lesion). The test does not differentiate first- from second-order neuron lesions in the setting of a positive dilation response. It should be noted that both cocaine and hydroxyamphetamine are difficult to obtain in many health care centres; a newer alternative involves administering apraclonidine drops, which produce pupil dilatation in patients with HS.

There is no specific treatment for HS; rather, therapeutic response is directed toward investigation and treatment of the underlying etiology. Generally this involves referral to an appropriate specialist as well as medical imaging.

Recommendations

The classic triad of HS is mild ptosis, miosis and anhidrosis of the head and neck. Careful clinical examination including testing of pupillary responses in light and dark conditions can help to differentiate among causes of anisocoria, such as HS, third-nerve palsy, Adie syndrome, and pharmacologic modification. Acquired HS in an adult raises the possibility of a head or neck lesion and must be investigated.

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Competing interests
None declared

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