Horner Syndrome

Horner syndrome, also called oculosympathetic paresis, is a neurological disorder in which the sympathetic arc that innervates structures in the head, face, and neck is disrupted. This can occur anywhere along the pathway from the hypothalamus to the ciliospinal center to the superior cervical ganglion to the ocular muscles. A classic triad of unilateral ptosis, anhidrosis, and miosis is highly suspicious for Horner syndrome. Diagnosis is clinical but MRI is often necessary to rule out structural lesions or find the underlying cause. Management involves treating the underlying cause.

PLAY PICMONIC

Pathophysiology

Sympathetic Disruption

Slipping Symba-lion

Horner syndrome can result from a lesion anywhere along a three-neuron sympathetic (adrenergic) pathway composed by first order neurons in the hypothalamus, preganglionic or second order neurons, and postganglionic or third order neurons that innervate the ipsilateral head, eye and neck.

Hypothalamus

Hippo-Thor with Symba-Neuron-leash

The posterolateral hypothalamus is home to first order neurons in the sympathetic arc. Central Horner syndrome can be caused be lesions within the hypothalamus or along the pathway from the hypothalamus to the preganglionic neurons in the ciliospinal center. Common lesions include tumors, stroke, and cervical spinal cord injury.

Ciliospinal Center

Seal with Spine-tail

The ciliospinal center is home to second order neurons in the sympathetic arc. It is located in the intermediolateral column between C8 and T2. From here, neurons project to stellate and superior cervical ganglion cells via the lung apices. This is why a superior lung tumor e.g. Pancoast tumor, can cause Horner syndrome.

Superior Cervical Ganglion

Super Cervical-cat and Gang-lion

After synapsing in the ciliospinal center, neurons project to the stellate and superior cervical ganglia where third order neurons are located. Postganglionic fibers travel from the superior cervical ganglion to muscles of the eye (sphincter pupillae, iris dilator) via the carotid plexus. As such, carotid artery dissection is a notable etiology of Horner syndrome.

Ocular Muscles

Eye Muscles

Ocular muscles like the iris dilator muscle fail to contract in Horner syndrome due to a lesion somewhere in the sympathetic pathway. This causes miosis and ptosis on sympathetic stimulation.

Clinical Triad

Ptosis

Toast-eyes

Ptosis occurs as a result of paralysis of the Müller's or superior tarsal muscle which has sympathetic innervation. It is a mild <2mm ptosis which is less severe than in cases of oculomotor nerve or elevator palpebrae lesions.

Anhidrosis

No-Sweatband

Anhidrosis on the face and arm will vary depending on the lesion. Postganglionic lesions usually don't present anhidrosis, however anhidrosis of the forehead could be seen. This sign is frequently not apparent to patients or clinicians.

Miosis

Mice-eyes

Ipsilateral miosis is more evident in dark than in light and dilatation of the affected pupil is slower by 15-20 seconds than normal.

Diagnosis

Diagnosis by Clinical Impression

Diagnostic-computer displaying Clinical Impression

If diagnosis is clear there is no need for diagnostic confirmation. However, in subtle cases confirmation is needed and can be done with cocaine or apraclonidine drops, the latter being more available. Optical administration of apraclonidine 0.5% would cause a reversal of anisocoria as Horner pupil will dilate by alpha-1 denervation supersensitivity while alpha-2 stimulation in the normal eye will cause slight constriction of the pupil.

MRI

M-R-eyes

Patients with Horner syndrome presentation require imaging, usually an MRI, unless it occurs in the setting of obvious trauma or after a surgical procedure to help identify the etiology of the disease.

Management

Treat Underlying Disorders

Treating Underlying Attacker

Treatment will depend on the underlying etiology, therefore cases of Horner syndrome with no identified etiology should be thoroughly investigated as there are life-threatening conditions that may cause this syndrome.