



Pupil disorder

Eye series – 19



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Case history

A 32 year old woman presents to the practice with a history of migraine. After a recent acute attack, the patient noticed that her right pupil was smaller than the left, being more noticeable in dim conditions. On the same side as the smaller pupil, the eyelid appears to have drooped slightly. She also notes that her forehead feels hot and flushed. There has been no change over the past week.

Question 1

List the differential diagnoses.

Question 2

Detail the sympathetic pathway to the eye.

Question 3

What conditions may lead to Horner's syndrome?

Question 4

What examination features, in particular of the pupil, would you look for in this patient?

Question 5

Is it possible to localise the cause of the pupil defect through patient signs or symptoms?

Question 6

What is the prognosis?

FEEDBACK

Answer 1

Differential diagnoses may include the following:

- direct ocular trauma may lead to an irregular pupil shape or function
- pharmacological side effects may lead to pupil dilation or constriction. Unless topical, the effect is usually bilateral
- complete third nerve palsy will result in a fixed and dilated pupil in the affected eye. Marked ptosis is usually present, which may mask a pupil defect. Concurrent extraocular muscle palsies will lead to motility disorders and diplopia. Third nerve palsies may also be pupil sparing. Causes may include vascular conditions (eg. aneurism), tumours and diabetes
- Horner's syndrome (*Figure 1*) is caused by an interruption of the sympathetic pathway to the eye. A common triad of symptoms are seen:
 - miosis of the ipsilateral pupil
 - a small to moderate ptosis in the upper lid (and a corresponding elevation of the lower lid [reverse ptosis]), and
 - anhydrosis of the ipsilateral side of the face.Both near and light pupil responses remain normal, however, there is usually a significantly slower dilation of the affected pupil in dim conditions (dilation lag). Horner's syndrome may be caused by various conditions such as tumours, trauma or migraine
- Adie's tonic pupil defect will cause the



Figure 1. Horner's syndrome

affected pupil to remain dilated. The pupil reacts poorly to light and this may increase the apparent difference between pupil sizes. A near reflex is usually present, although this may be reduced. Re-dilation of the pupil will also be slower in the affected pupil. Reduced or loss of accommodation may occur and the patient may complain of blurred vision with near work which may resolve over time. Long term, the affected pupil will constrict in size and remain fixed. Usually benign in origin, it typically affects women in their 30–40s. It may follow a viral illness

- Argyll Robertson pupil defect usually presents with small, irregular pupils that are sluggish or nonresponsive to light, similarly the pupils will dilate poorly with dim or pharmacologically induced conditions. The pupils will respond promptly to a near response (accommodation). Usually caused by neurosyphilis, this condition is almost always bilateral in presentation.

Answer 2

The sympathetic pathway to the eye has three distinct sections. The first section (first order neurons) originates in the hypothalamus. The fibres descend to the level of the eighth cervical and fourth thoracic vertebrae of the spinal cord where the fibres then synapse with second order preganglionic fibres.

Second order fibres pass over the apex of the lung and enter the superior cervical ganglion in the neck synapsing with postganglionic fibres. These third order neurons travel via the internal carotid artery and enter the eye through the cavernous sinus. Fibres then branch to innervate the dilator muscle of the iris and the muscle of Mueller within the eyelid (responsible for initiating eyelid retraction during eyelid opening). Travelling via the external carotid artery the postganglionic fibres also serve to innervate the sweat glands of the face.

Answer 3

Interruption of the pathway at any point along the sympathetic pathway will lead to the classic Horner's triumvirate of symptoms: miosis, mild ptosis and anhydrosis. Conditions leading to Horner's syndrome include:

- trauma to the head or neck region (including surgical trauma)
- aneurysms of the aorta or carotid arteries
- vascular disease (eg. brain stem stroke)
- demyelination disorders (eg. multiple sclerosis)
- malignancy of the lungs (eg. Pancoast's tumour) and neck regions (eg. Hodgkin disease).

A careful history and examination is required to establish the underlying cause of disease in a patient with Horner's syndrome.

Answer 4

Comparing the lid position of both eyes in primary position will provide evidence of ptosis. Look for an area of dry, hot skin along the ipsilateral facial and/or neck region (reduced innervation of the sweat glands with Horner's syndrome) and perform a neu-

rological examination looking for weakness of limbs or sensory loss that may occur in combination with the Horner's syndrome.

A controlled pupil examination will generally provide the necessary information to classify the defect. The practitioner should begin with an examination of the eye for local causes of the pupil irregularities such as iris trauma or inflammation.

The next step is to determine the response to light and dark conditions. If the pupil difference (anisocoria) is greater in the dark, it usually represents a dilation defect, ie. Argyll Robertson or Horner's defect. If anisocoria is greater in light conditions, a problem with constriction is more likely, ie. Adie's tonic pupil. To further differentiate these conditions the response to near stimulus is necessary. Adie's pupil will react sluggishly to a near stimulus. If there is no response to either light or near conditions, a third nerve lesion or pharmacological condition may exist.

A 'swinging flashlight' test is used to detect a relative afferent defect of the optic nerve. Each pupil is stimulated with light in quick succession. The abnormal pupil will dilate instead of constricting when stimulated. This is because the corresponding action of dilation of the normal eye (when the light source is removed) will be stronger than the constriction effect caused by the light in front of the affected eye.

Answer 5

In many cases the astute observer will be able to localise the cause of Horner's syndrome by the patient's clinical signs. Gross neurological deficits such as hemiparesis or analgesia, dysarthria, ataxia and loss of sweating of the entire half of the body combined with Horner's pupillary defects may indicate lesions of the first order neurons. Second order neuron lesions are commonly associated with trauma to the chest and neck. The patient may complain of facial, neck or chest pain. This may also result from compression of the fibres due to an apical lung tumour (Pancoast's tumour). Horner's syndrome in the presence of acute

onset facial or neck pain without a history of trauma may indicate carotid artery dissection from pre-existing cardiovascular disease. Anhydrosis of the face and neck is also suggestive of second order involvement.

Third order or postganglionic lesions are commonly vascular in origin and usually occur in isolation of neurological deficits, although the patient may suffer from facial pain or ear, nose and throat disease. Anhydrosis is generally limited to the ipsilateral forehead.

Answer 6

In the majority of cases, Horner's syndrome is permanent, although transient episodes may be seen in cases following cluster migraine attacks. Rate of resolution of the syndrome, if it does occur, may vary considerably. The main goal is to detect and treat the underlying cause. Recognising the presence of Horner's syndrome and immediate referral to an appropriate specialist is paramount to the patient's health.

AFF

Conflict of interest: none declared.

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The 5 domains of general practice



1. Communication skills and the patient-doctor relationship



2. Applied professional knowledge and skills



3. Population health and the context of general practice



4. Professional and ethical role



5. Organisational and legal dimensions