Unilateral erythema or unilateral pallor?

IMAGES IN MEDICINE

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The photograph shows miosis, ptosis and hemifacial anhidrosis with pallor on the left side in a previously healthy woman in her thirties. The photograph and video were taken following an exercise session.
The patient was referred to a neurologist due to a two-month history of unilateral facial flushing. Clinical examination revealed mild miosis, ptosis, anhidrosis and pallor on the left side of the face, with a sharp midline border. She had no other focal neurological deficits. Activity would cause her right cheek to redden, but not her left. A thorough work-up with repeated clinical testing and imaging (contrast-enhanced MRI of the brain, neck, brachial plexus, and the apical zone of the lungs with angiography) revealed no lesions in the oculosympathetic pathway or other precipitating factors. Clinical findings were consistent with left-sided Horner’s syndrome.

The oculosympathetic pathway begins centrally with the first-order neuron in the hypothalamus and ends in the spinal cord (C8–Th3). A preganglionic (second-order) neuron continues from here, terminating in the superior cervical ganglion, where it forms synapses with several postganglionic (third-order) neurons. These in turn project to the pupil (1).

Unilateral Horner’s syndrome can result from various aetiologies, both benign and malignant, and with varying localisation. In up to one-third of patients, the aetiology remains unknown (2). The incidence of unilateral Horner’s syndrome is approximately 2%, and symptoms resolve in more than half of patients (3).

When the first-order neuron is involved (most often due to an infarct in the lateral medulla oblongata), anhidrosis is usually seen in the head, neck, arm and upper body on the affected side. However, the reduction in sweating is often overshadowed by other signs and symptoms from the central nervous system. Involvement of preganglionic fibres (typically as a result of a tumour near the apex of the lung) can give rise to Horner’s syndrome with varying degrees of ipsilateral anhidrosis, whereas involvement of third-order neurons (usually due to dissection of the internal carotid artery) rarely leads to anhidrosis. This is because vaso- and sudomotor fibres to the face follow the external carotid artery. Most cases of isolated Horner’s syndrome are caused by a lesion in a third-order neuron.
The difference in the anisocoria is more pronounced – or more visible – in dim lighting and results from interruption of the nerve supply to the iris dilator muscle, whereas ptosis results from interruption of the nerve supply to the superior tarsal muscle. Interrupted sympathetic innervation of the facial blood vessels and sweat glands can give rise to erythema as a result of vasodilation, and to dry skin because of reduced sweat secretion. However, the denervation makes the blood vessels supersensitive to circulating adrenaline, and therefore the side of the face with the sympathetic defect is usually palest, as can be seen in the video.

The patient has consented to the publication of this article. With thanks to Sindre Bondi for assistance with recording the video and processing the image.

LITERATURE

1. Alstadhaug KB. Ervervet Horners syndrom. Tidsskr Nor Legeforen 2011; 131: 950–4. [PubMed] [CrossRef]
