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What can a thermal imaging camera tell you? Idiopathic Harlequin Syndrome

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To the Editors,

A 50-year-old male patient with suspected tension-type headache was admitted to the neurology department for further diagnostics. The patient reported that symptoms had begun c.4 years ago, without visual disturbances, nausea, vomiting, and tearing or redness of the conjunctiva during pain.

Additionally, the patient ocasionally experienced episodes of hemi-cooling, pallor and decreased sweating on the left side of his face during intense physical exercise. He stated that the right side of his face was red, properly insulated and sweated normally. During one of the episodes, the patient took a photo at work with a thermal imaging camera (just for 'fun'), which showed a significant temperature difference on either side of his face: c. 36.8 °C on the right side and 32.6 °C on the left side (Fig. 1).

The neurological examination showed no abnormalities.

Harlequin Syndrome was suspected. Differential diagnosis was performed, including MRI of the head and cervical spine, lung X-ray and Doppler ultrasound of the carotid arteries; secondary causes of damage to the sympathetic nervous system were excluded.

Idiopathic Harlequin Syndrome was diagnosed, and the patient was informed about the mild nature of the disease and the factors that could trigger episodes of the disease.

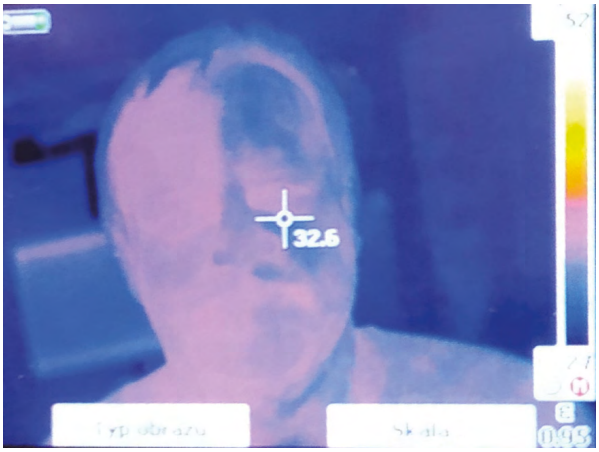

Figure 1. Photo taken by thermal imaging camera illustrating different temperatures in either half of face

Harlequin Syndrome (HS) is a rare disorder of the autonomic system caused by unilateral damage to sympathetic fibres originating from the Th2–Th3 level of the spinal cord which are responsible for the vasomotor and sudomotor innervation of the ipsilateral half of the face. It was first described in 1988 by Lance et al. HS more often affects women than men. Symptoms appear suddenly after physical exercise, strong emotions or eating a spicy meal and are characterized by hemifacial discolouration sharply demarcated at the midline. On the side of the

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Table 1. Causes of secondary Harlequin Syndrome [1–5]

Organic
Pancoast tumour
Thoracic syrinx
Dissection of cervical carotid artery
Cervical sympathetic tumours
Medullary infarction
Thalamic haemorrhage
Birth injuries
Guillain-Barre Syndrome
Diabetic polyneuropathy
Multisystem atrophy
Pure autonomic failure
CMV or HSV-1 infections
Iatrogenic
Paravertebral blocks
Intrascalene block
Thoracic epidural
Erector spinae plane block
Thyroidectomy
Carotid artery endarterectomy
Anterior cervical discectomy
Thoracic sympathectomy
Mediastinal or cervical mass excision

autonomic fibre damage, the face is pale, cool and with decreased sweating, while on the opposite side it is red, normally warm and with normal or increased sweating. Symptoms last from several minutes to several hours, and disappear spontaneously [1–5].

Sometimes there is a subclinical form of HS, in which there are only differences in temperature on both sides of the face without a change in colour. In more than 50% of cases, the disease is idiopathic. It is worth mentioning that HS is common among newborns due to the immaturity of hypothalamic and vasomotor functions. Secondary Harlequin Syndrome can have many causes (Tab. 1) [4].

Making a diagnosis requires a thorough differential diagnosis (imaging of the brain, cervical spinal cord, lungs, carotid arteries) aimed at excluding secondary causes [2, 4, 5].

Idiopathic Harlequin Syndrome is mild and does not require any treatment. If the symptoms are uncomfortable for the patient, treatment with injections of botulinum toxin or stellate ganglion blockade may be attempted. In a case of the secondary form, treatment is aimed at removing the cause that is provoking the condition [1, 4].

Knowledge of Harlequin Syndrome symptoms allows a quick diagnosis to be made while excluding secondary causes, and protects the patient from unnecessary tests and therapeutic interventions.

Article information

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