



HYPERHIDROSIS

TWO FACES: WHAT ABOUT HARLEQUIN SYNDROME?

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Background: Harlequin syndrome (HS) is a rare clinical condition characterized by a unilateral erythrosis of the face with hyperhidrosis and contralaterally a pale anhydrotic aspect. It is mainly idiopathic but it can be associated with severe diseases. Herein, we report two patients with HS.

Observation:

Observation N° 1: A 27-year-old man with a history of migraine, consulted us for a unilateral erythrosis on his face. He reported a redness associated with a hyperhidrosis with strictly medial limits at the left side. It contrasted with anhidrosis and a normal appearance of contralateral hemiface. These attacks were triggered by exercise. They spontaneously disappeared at rest. The clinical examination especially neurological assessment showed no abnormality.

Observation N° 2: A 22-year-old man with no special medical history. He complained about same symptoms as the first patient but the attacks occurred without any triggering factor. The neurological examination revealed an incomplete Horner's syndrome (eyelid ptosis, enophthalmos and a tendency to miosis).

The two patients were diagnosed with HS. In the two cases, the etiological investigation was normal.

Key message: HS corresponds to a unilateral dysfunction of sympathetic system characterized by hypohidrosis and loss of facial erythrosis to heat, exercise, or emotional factors. This phenomenon is compensated by excessive contralateral sweating and redness. It may be isolated or integrated into other dysautonomic syndromes like Horner's syndrome. In most cases, this syndrome is idiopathic. However, in some cases, this syndrome resulted from compressive organic denervation of the T2 and T3 roots. Concerning treatment, a contralateral sympathectomy can be proposed but it may be at risk of many complications. Recently, the injections of botulinum toxins seems to be a promising therapeutic alternative. Despite the rarity of HS, dermatologists should be aware of this clinical condition in order to diagnose properly and eliminate a compressive lesion.

