Harlequin syndrome in childhood - Case report
Síndrome de Arlequim na infância - Relato de caso

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Abstract: Harlequin syndrome happens in only one side of the face. In the affected half, the face does not sweat or flush even with simulation. Arms and trunk could also be affected. This condition is induced by heat, exercise and emotional factors. The article reports a case of a nine-year-old female with a 3-year history of unilateral flushing and sweating after exercise; a brief literature review is included. Despite the rarity of this syndrome, dermatologists should recognize this condition and refer these patients to ophthalmological and neurological examination.

Keywords: Erythema; Hypohidrosis; Pediatrics; Syndrome

INTRODUCTION

Harlequin syndrome is characterized by unilateral diminished facial flushing and sweating in response to heat, exercise or emotional factors.

The entity was first described by Lance and Drummond in 1988, who proposed the radicular arterial occlusion, at the third thoracic section, as the pathogenic mechanism. This lesion may involve sympathetic fibers as well as parasympathetic neurons of the posterior and ciliary ganglia. 1

Lance et al. named it Harlequin syndrome discolouration based on the classical Italian theatre character from “Commedia dell’Arte”. 2

More and more evidence, as the first Harlequin case, indicates that this dysautonomia is not always confined to facial skin, being even more diffuse and extensive than the previous concept. According to Cheshire and cols.(2008), some cases of Harlequin syndrome may have irregular and diffuse distribution, combining sympathetic and parasympathetic lesions, with ipsilateral face and arm anhidrosis and absence of flushing. 2

CASE REPORT

A nine-year-old female patient reported unilateral flushing associated with ipsilateral sweating when submitted to prolonged physical exercise. It started at the age of six, being unaltered since then. The opposite side of the face remains pale and anhidrotic.
These skin changes could be reproduced during her examination, 30 minutes after exercising, with flushing and sweating (Figure 1). In contrast to the patient’s beliefs, the affected side is not the one with normal vascular and sweat response, but the pale and anhidrotic side. Her medical history had no other diseases or accidents. No birth problems (such as forcepts usage) were identified. A neurological and ophthalmological evaluation made by a neuropediatrician and an ophthalmologist was normal (with no need for additional tests).

DISCUSSION

Harlequin syndrome is a term reserved for patients with an autonomic disturbance that leads to unilateral anhidrosis and reduced or absent facial flushing. It is a rare but remarkable disease. 

Oculosympathetic paresis may be associated but is not always present and not evident in our patient.

Lance and cols. also raised the theory that the contralateral side may present an increase in sympathetic activity, with hyperhidrosis. That would justify the fact that four out of five reported patients had transpiration triggered by gustative stimuli. The secondary abnormality would be an effort to compensate the deficient side.

Women may be more affected than men, however this could be due to the higher social prejudice against asymmetry in this group. Breunig and Smith observed, in their study of 39 patients with this condition, a mean age of 47 years for men and 45 for women. Most of the cases reported in literature are of benign nature. It is predominantly idiopathic (as in our patient), but may be associated with brainstem infarct, superior mediastinum neurinoma, internal jugular vein catheterism or spinal invasion of the left apical lung cancer. Images should be performed according to symptoms and medical history.

Sarikaya and cols. suggested the spontaneous carotid dissection as a possible etiology for Harlequin syndrome, when reporting a case of a 52-year-old man who, after suffering this particular vascular accident, started showing the symptoms when practicing exercises. Darvall et al reported Harlequin syndrome in a 26-month-old baby, submitted to an elective procedure for emptying a left cervical lymphatic malformation.

It can also result from regional infection by a neurotropic virus. Pathogenic viruses may have preference for the stellate ganglion, as the herpes simplex virus type 1 does through the geniculate ganglion in Bell’s palsy, and as the cytomegalovirus does in Ross syndrome.

The dysfunction seen in this disease occurs by an alteration of sympathetic nervous system activity, secondary to the lesion of the preganglionic fibers, superior cervical ganglion or postganglionic fibers, in the external carotid plexus.

Occasionally, Harlequin syndrome may be superimposed by other syndromes. Bremner and Smith found, in concordance with the literature, that most patients have normal pupils (as our patient). However, the abnormality more frequently found in their patients (46%) is Horner syndrome, characterized by ptosis, miosis, and enophthalmus.

Harlequin syndrome patients may also present symptoms that are seen at Adie syndrome (tonic pupils) and at Ross syndrome (tonic pupils and segmental anhidrosis).

The clinical scene reported in these conditions indicate a major autonomic deficit involving other parts of the nervous system, including parasympathetic lesion. This point may be considered for the differential diagnosis.

Alternatively, Harlequin syndrome may simply
be a common dysautonomic manifestation of many diseases, which asymmetrically affect sympathetic vasomotor innervations, including the Guillain-Barre syndrome, Pure Autonomic Failure, Multiple System Atrophy and Diabetic Neuropathy. Unilateral facial flushing may also be the first symptom of more serious diseases, as neoplasms or cerebral vascular accident; therefore, a particular investigation is indicated, if there are any suspicions. Based on the sympathetic anatomy, investigation techniques may reveal standards of symptoms that indicate the site of the lesion. Imaging techniques, as magnetic resonance and ultrasound, and electrophysiology, may be done to find the underlying cause.

Most of the Harlequin syndrome cases do not require medical treatment, unless there is an underlying disease. Thus, it is necessary to explain the disease physiopathology and its favorable prognosis to the patient. For patients with serious social embarrassment due to unilateral flushing, there is the option of contralateral sympathectomy. In this procedure, the flushing on the normal functioning side of the face is interrupted. However, this procedure must have restricted indication, since it is neurodestructive.

Dermatologists should recognize this condition and refer these patients to neurological and ophthalmological examination.

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