HARLEQUIN SYNDROME – A RARE ENTITY

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ABSTRACT
Harlequin syndrome is a condition which affects one half of the face. In the affected half, the face does not sweat or flush even with simulation. The condition may also affect arms and trunk. This condition is induced by heat, exercise and emotional factors. Despite the rarity of this syndrome, it should be recognized and evaluated for the associated ophthalmologic and neurologic complications.

Keywords: Harlequin Syndrome, Facial Flushing, Neurology, Ophthalmologic

INTRODUCTION
Harlequin syndrome is characterized by unilateral diminished facial flushing and sweating in response to heat, exercise or emotional factors. It was first described by Lance et al., in 1988. He theorized that the radicular artery occlusion, at the third thoracic section, is the etiology. The lesion may involve sympathetic fibres as well as parasympathetic neurons of the posterior and ciliary ganglia. Lance et al., (1988) named it Harlequin syndrome discoloration based on the classical Italian theatre character from "Commedia dell'Arte".

CASES
A 28 years old female patient presented to us with complaints of unilateral right sided flushing associated with ipsilateral sweating involving the face and upper chest when submitted to prolonged physical exercise or exposed to sun. She noticed this since she was seven years old. The opposite side of the face remains pale and anhidrotic. These skin were reproduced after 30 minutes after exercising, with flushing and sweating (Figure 1). She always thought that the right side is pathologic side. Her past medical history was insignificant. No birth problems (such as forceps usage) were identified. In general examination Pulse was 76/min, regular. BP was 118/74 mm of Hg. There was no postural fall. In neurological examination higher functions were normal. In cranial nerve examination 2nd cranial nerve was normal (pupil sizes, light reflex and accommodation reactions), all other cranial nerves were normal. Motor system examination was normal, DTR were normal and there was no sensory loss. Cardiovascular system was normal.

Investigations: Complete blood count, Fasting and post meal blood sugar, Liver and Kidney functions were normal. EMG/NCV was normal. Thyroid profile, serum uric acid levels were normal. CXR chest and carotid Doppler was normal. A diagnosis of Harlequin Syndrome was made.

DISCUSSION
Harlequin syndrome is a rare autonomic disturbance that leads to unilateral anhidrosis and reduced or absent facial flushing (Moon et al., 2005; Cheshire and Jr Low, 2008). 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. Oculosympathetic paresis may be associated (Cheshire and Jr Low, 2008). It is also hypothesized that the contralateral side may present an increase in sympathetic activity, with compensatory hyperhidrosis (Tascilar et al., 2007).
Case Report

Women may be more affected than men. In a 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). It can be associated with brainstem infarct, superior mediastinum neurinoma, internal jugular vein catheterisation, spontaneous carotid dissection and spinal invasion of the left apical lung cancer (Tascilar et al., 2007). Images should be performed according to symptoms and medical history. Traumatic cervical lesions were also reported as a cause of this syndrome, secondary to a rupture of the vasomotor neurons located in the sympathetic cervical chain (Darvall et al., 2008). Rarely is can be associated with Horner’s syndrome (ptosis, miosis, anhidrosis and enophthalmus) and Ross syndrome (tonic pupils and segmental anhidrosis) (Caparros-Lefebvre et al., 1993).

Other disorders with dysautonomia like Guillain-Barre syndrome, Pure Autonomic Failure, Multiple System Atrophy and Diabetic Neuropathy should be ruled out by imaging techniques, as MRI and electrophysiology (Wasner et al., 2005).

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.

REFERENCES


Case Report

