Harlequin syndrome : Double face

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Abstract

Harlequin syndrome corresponds to unilateral dysfunction of sympathetic system, characterized by flush and unilateral hyperhidrosis associated with hypo or anhidrosis and paleness of the opposite side. It is, usually, idiopathic. Rarely, it may be associated with compressive organic processes, iatrogenic causes and general diseases. It is a real therapeutic challenge.

Title page

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A written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy"

A key clinical message:

Harlequin syndrome corresponds to unilateral dysfunction of sympathetic system, characterized by flush and unilateral hyperhidrosis associated with hypo or anhidrosis and paleness of the opposite side. It is, usually, idiopathic. Rarely, it may be associated with compressive organic processes, iatrogenic causes and general diseases. It is a real therapeutic challenge.

The Harlequin syndrome, was described for the first time by Lance et al in 1988, as an uncommon disorder of the sympathetic nervous system [1]. It is characterized by unilateral diminished sweating and heat- or exercised-induced facial flushing [1]. We present the cases of two patients with this rare syndrome, including a review of the literature.

Patient N°1:

A 27-year-old man, with a medical history of migraine, consulted for flushing and an excessive sweating on the left side of his face evolving for few months. This contrasted with anhidrosis and normal appearance of the right side of his face (Fig. 1). These skin changes were triggered by physical exercise and disappeared spontaneously at rest. Dermatological examination at rest revealed no abnormalities. Findings of physical and neurological examinations were normal. Routine laboratory studies and carotid artery ultrasonography yielded normal results. Thus, the diagnosis of idiopathic Harlequin was established.

Patient N°2:

A 22-year-old man, with no specific medical history, presented with an excessive sweating and flushing on the left side of his body including his face evolving by relapses for 2 years. No triggering factor was found. On examination at rest, we noted an asymmetric flushing and hyperhidrosis limited to the left side of his face, neck and upper trunk (Fig. 2). The neurological examination revealed a left ptosis, a pupillary asymmetry with a tendency to miosis, leading to an incomplete Claude Bernard Horner syndrome (CBH). Bilateral pupillary paresis was also noted. The blood pressure was within the normal range. Routine laboratory studies, carotid artery ultrasonography, magnetic resonance of angiography of the head ruled out secondary causes. So, we evoked the diagnosis of idiopathic Harlequin syndrome associated with CBH.

To the best of our knowledge, only almost one hundred cases have been reported through literature [2]. This phenomenon corresponds to unilateral dysfunction of the sympathetic system [2, 3]. It is characterized by flush with unilateral hyperhidrosis associated with hypo or anhidrosis and paleness of the opposite side [1-3]. This was explained by a compensatory mechanism on the unaffected sympathetic innervated side [1-3]. Arms and trunk may be affected, as well as our patient [4, 5]. Some triggering factors were reported, such as: heat, emotions or physical efforts, which are the usual stimuli of the sympathetic nervous system [1-3]. This phenomenon was reported in association with dysautonomic syndromes such as CBH, as illustrated in our second patient [3]. The association between Harlequin syndrome and migraine was also described, as well as our first patient [1, 2]. In most cases, Harlequin syndrome is idiopathic [1-3]. However, rarely this syndrome may be secondary to compressive organic processes of T2 and T3 sympathetic trunks such as medullary astrocytoma, mediastinal neuroma, pulmonary neoplasia of the apex, or carotid dissection [1-8]. Iatrogenic causes are increasingly described in particular jugular venous catheterization, thoracic sympathectomy, or after resection of anterior mediastinal tumor [1-8]. In addition, some general diseases may also cause Harlequin syndrome, including Barred Guillain syndrome, diabetic neuropathy, or multiple sclerosis [1-8]. The diagnostic approach seeking systematic secondary etiologies of Harlequin syndrome requires a rigorous interrogation. It is recommended to study the carotid cervical arterial by carotid artery ultrasonography network and even magnetic resonance of angiography of the head [2, 4]. The phenomenon of Harlequin is most common in women patients at the third decade leading to an important social embarrassment requiring treatment [2]. Apart from the secondary cases where the compressive origin can be surgically removed, the therapeutic possibilities are very few; limited to contralateral sympathectomy, repeated block of the stellate ganglion and botulinum toxin [9, 10].

Through our case report, we would like to make clinician aware of this rare syndrome and its management options in order to ameliorate quality of life among patients suffering from Harlequin syndrome.

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Figures:

Figure 1: A flushing and an excessive sweating on the left side of the face contrasting with

anhidrosis and normal appearance of the right side.

Figure 2: An asymmetric flushing and hyperhidrosis limited to the left side of the face.

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