INTRODUCTION

The Harlequin syndrome was described for the first time by Lance et al in 1988, as an uncommon disorder of the sympathetic nervous system. With only one hundred cases reported, it is characterized by unilateral facial flushing and 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.

CASE PRESENTATION

2.1 Patient number 1

A 27-year-old man, with a medical history of migraine, consulted for flushing and 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. This skin changes were triggered by physical exercise and disappeared spontaneously at
rest. Dermatological examination at rest revealed no abnormalities. The 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. We opted for botulinum toxin. The patient did not accept, as for him, it was a very expensive treatment.

2.2 | Patient number 2

A 22-year-old man, with no specific medical history, presented with 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 (Figure 2). Neurological examination revealed left-sided ptosis, tendency to miosis, and minimal pupillary response to light and cocaine eye drops without relative afferent pupillary defect, leading to the diagnosis of 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, and magnetic resonance angiography of the head ruled out secondary causes. So, we evoked the diagnosis of Harlequin syndrome associated with CBH. Botulinum toxin was proposed to him, but the patient refused the treatment, as it was very expensive.

3 | DISCUSSION

To the best of our knowledge, almost one hundred cases of Harlequin syndrome have been reported through literature. The phenomenon of Harlequin is most common in women patients in the third decade. Only 37 cases of pediatric Harlequin syndrome have been described. This phenomenon corresponds to unilateral dysfunction of the sympathetic system. It is characterized by a flush with unilateral hyperhidrosis associated with hypo or anhidrosis and paleness of the opposite side. This was explained by a compensatory mechanism on the unaffected sympathetic innervated side. The arms and trunk may be affected, and our second patient. Some triggering factors were reported, such as heat, emotions, or physical efforts, which are the usual stimuli of the sympathetic nervous system. This phenomenon was reported in association with dysautonomic syndromes such as CBH, as illustrated in our second patient. The association between Harlequin syndrome and migraine was also described, and our first patient. In most cases, Harlequin syndrome is idiopathic. 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. Iatrogenic causes are increasingly
described such as Harlequin syndrome following jugular venous catheterization, thoracic sympathectomy, or after resection of anterior mediastinal tumor.\(^1\)\(^9\) In addition, some general diseases may also cause Harlequin syndrome, including Guillain Barre syndrome, diabetic neuropathy, or multiple sclerosis.\(^1\)\(^9\) The diagnostic approach seeking secondary etiologies of Harlequin syndrome requires rigorous clinical examination. It is recommended to study the carotid cervical artery by ultrasonography and even magnetic resonance angiography of the head.\(^2\)\(^5\) Finally, Harlequin syndrome may lead to 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 injection of botulinum toxin.\(^10\)\(^11\)

4 | CONCLUSION

Through our two reported cases, we would like to make clinicians aware of this rare syndrome and its management options in order to ameliorate the quality of life among patients suffering from Harlequin syndrome.

AUTHOR CONTRIBUTIONS

Korbi Mouna involved in conception, exploration, redaction, and revision. Boumaiza Sirine involved in redaction and revision. Achour Asma involved in exploration. Belhadjali Hichem involved in validation and conception. Zili Jameleddine involved in validation.

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None.

CONFLICT OF INTEREST

None.

ETHICAL APPROVAL

None.

CONSENT

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

DATA AVAILABILITY STATEMENT

None.

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