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(CASE REPORT)

Idiopathic Harlequin syndrome: A case report and literature review

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Abstract

Harlequin's syndrome is a rare dysautonomic syndrome of the face characterized by sweating with flush of one side and anhidrosis of the contralateral side. Mostly idiopathic although several secondary cases have been reported in the literature, the purpose of the treatment is mainly aesthetic and functional. We report the case of a patient having harlequin syndrome in its idiopathic form with a literature review.

Key words: Harlequin Syndrome; Sympathetic System; Anidrosis; Idiopathic; Treatment

1. 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 [1]. it is characterized by unilateral facial flushing and hyperhidrosis associated with hypo or anhidrosis and paleness of the opposite side. It is often idiopathic, but it may be associated with compressive organic processes, iatrogenic causes, and general diseases [1]. In this article, we report a case of idiopathic Harlequin syndrome and review the literature.

2. Case Report

Our patient is 40 years old, having as antecedents an essential epilepsy, Elsewhere, the patient has no history of diabetes or neuropathy, no previous cervicothoracic surgery or arterial catheterization, no history of trauma. He consulted for 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 disorder was noted since childhood, during sports efforts and more frequently in the hot season, otherwise the patient does not report other signs including ocular or neurological. The dermatological examination did not show abnormalities, no asymmetry of the face. Her neurological examinations including higher mental function, cranial nerves, motor and sensory systems, deep tendon reflexes, and coordination were all normal. Specifically, there were no pupillary abnormalities, including Horner syndrome. The patient was instructed to exercise for more than an hour. Examination immediately after exercise revealed unilateral excessive sweating on the left side contrasted with anhidrosis and normal appearance of the right side of his face.(Figures 1 and 2) Magnetic resonance imaging (MRI) of the brain and cervicothoracic spine was normal. Computerized tomography (CT) scan of the chest, including the area of the thoracic sympathetic chain, was normal. The diagnosis of the harlequin syndrome was then retained.

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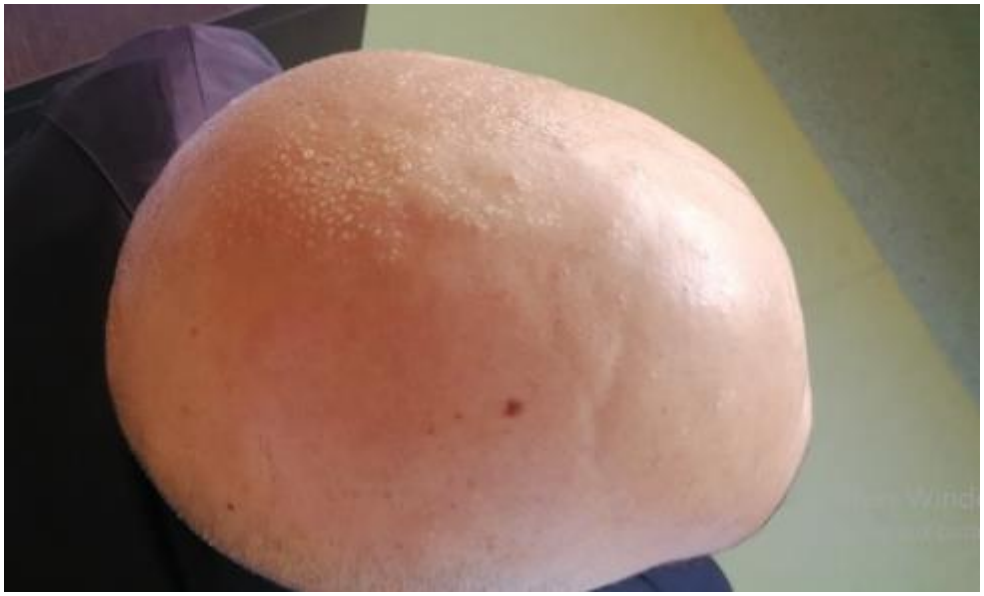

Figure 1 Excessive sweating on the left side of head contrasted with anhidrosis and normal appearance of the right side of head

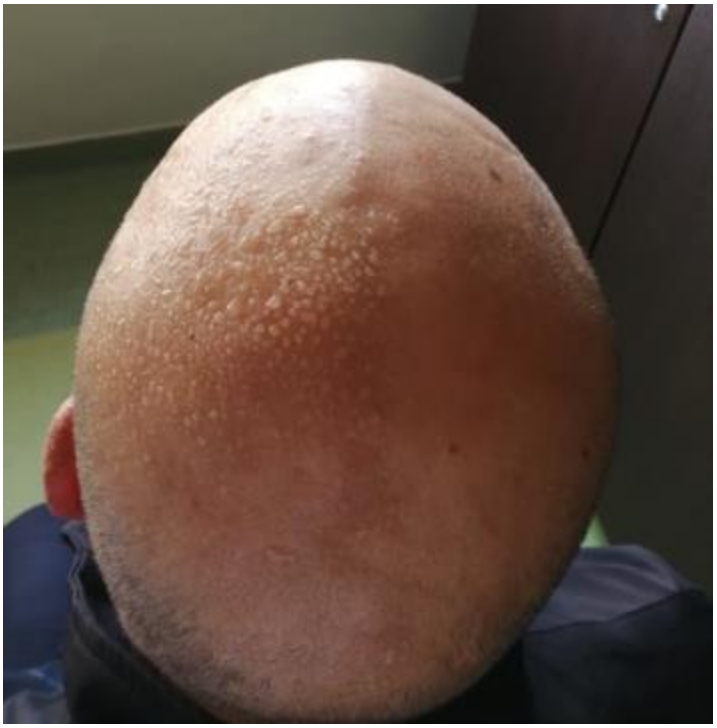

Figure 2 Excessive sweating on the left side of head contrasted with anhidrosis and normal appearance of the right side of head

3. Discussion

The harlequin syndrome is the expression of an abnormality of the sympathetic nervous system, originating from the T2-T3 roots or their nerve fibers, the site of the lesions may be at the level of sympathetic and parasympathetic nerve fibers of the stellate ganglion. It is a unilateral blocking of sympathetic innervation with absence of cutaneous vasodilatation and sweat secretion in response to a thermal, emotional or other stimulus [2]. This causes compensatory contralateral sympathetic hyperstimulation, explaining hypersudation with a flush. This syndrome is triggered most often after exercise and following exposure to heat, as reported by our patient. Clinically, it corresponds to a unilateral

facial erythrema with flush and hypersudation [6] giving the appearance of a harlequin face reminiscent of the black red mask of Arlequin, well known in European folklore [3].

Harlequin syndrome is usually idiopathic but could be the first manifestation of several disorders such as Guillain-Barre syndrome, Bradbury-Eggleston syndrome, and diabetic neuropathy. The syndrome also might be caused by brainstem infarction, carotid artery dissection, toxic goiter, superior mediastinal neurinoma, syringomyelia, multiple sclerosis, internal jugular vein catheterization, and iatrogenic effects of invasive procedures [4]. It is usually acquired but can be congenital in up to 6% of the reported cases [5]. 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 [6,7]. As it can remain most often idiopathic and that was the case of our patient. Indeed, on a review of literature, published by Guillotton et al., 59 out of 108 cases were idiopathic [6]. The spontaneous regression of this primitive syndrome has not been reported and studies are evolving in order to better understand its physiopathology [10].

Finally, Harlequin syndrome may lead to important social embarrassment requiring treatment [6]. 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 [8,9].

4. Conclusion

Harlequin syndrome corresponds to unilateral dysfunction of the 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. Medical or surgical treatments are not required for idiopathic Harlequin syndrome, but social and psychological factors may indicate sympathectomy or botulinum toxin injection.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no conflicts of interest.

Statement of ethical approval

The authors declare that that the study complies with the ethical requirements of Unversity of Hassan II Casablanca.The Ethics Committee of has confirmed that ethical approval is not required for a case report.

Statement of informed consent

The author has received the written informed consent from the patient for publication of this case report.

Authors' Contributions

- Mohamed Amine Mnaili, MD, contributed to the content, references, and written manuscript of the case report.
- Ahmed Bourazza, MD, contributed to expert revisions, details, and references of the manuscript.

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Data availability

The data that support the findings of this report are available from the corresponding author, upon reasonable request.

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