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Case Report

Idiopathic Harlequin syndrome: a rare presentation of unilateral anhidrosis

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ABSTRACT

Harlequin syndrome is a rare disorder of sympathetic dysfunction characterized by unilateral anhidrosis or hypohidrosis with contralateral flushing and compensatory sweating. Approximately 100–125 cases have been documented in the literature, with the majority involving the face. Diagnosis is primarily clinical and requires exclusion of secondary neurological or structural causes. Herein, we report a male patient in his 40s presenting with anhidrosis over the right half of the trunk for the past 8 years, associated with occasional contralateral flushing and no facial involvement. Neurological and ophthalmological examinations were unremarkable. MRI of the cervical spine revealed mild disc bulges without clinically significant cord pathology explaining the symptoms. Following clinical evaluation and exclusion of relevant secondary causes, a diagnosis of idiopathic Harlequin syndrome was considered. This case highlights the uncommon presentation of isolated truncal involvement and the importance of clinical recognition.

Keywords: Harlequin syndrome, Anhidrosis, Hypohidrosis

INTRODUCTION

Harlequin syndrome was first described by Lance et al in 1988.[1] It is a condition in which there is sympathetic dysfunction characterized by unilateral hyperhidrosis and flushing associated with hypohidrosis and xerosis on the contralateral side. It can occur due to compressive causes but the most common cause is idiopathic.[2] The condition typically becomes more noticeable during physical exertion, emotional stress or exposure to heat. Diagnosis is primarily clinical, although further investigations may be required to exclude underlying structural or neurological abnormalities. This syndrome is a therapeutic challenge with limited treatment options for the treating clinician.

CASE REPORT

A male patient in his 40s, hailing from the central part of India, presented with complaints of absent sweating over the right half of the trunk region for the last 8 years, while sweating remained normal on the left side, with occasional episodes of flushing. The symptoms had persisted over this prolonged duration without any other associated neurological complaints or significant systemic manifestations. There was no history of trauma. No surgical history. No history of stroke or cardiac disease.

The patient had no other neurological complaints; pupillary size and reactivity, as well as deep tendon reflexes, were normal, with no evidence of facial involvement, orthostatic hypotension, or sympathetic dysfunction elsewhere on the body. No familial occurrence or hereditary pattern of similar symptoms was identified. No history of any significant comorbidities.

The clinical history was predominantly characterized by the long-standing unilateral absence of sweating, with preservation of normal sweating on the contralateral side.

On examination, multiple dry xerotic patches were seen on the right side of back and chest while the left side appeared normal. Mild flushing was noted on the upper aspect of the left side of the back (Figure 1).

Routine laboratory investigations like complete blood count, liver function test, serum creatinine, serum TSH levels, and serum IgE levels were done as part of the initial diagnostic evaluation and were within normal limits. Magnetic Resonance Imaging (MRI) of the neck was done and revealed a mild disc bulge with small posterocentral protrusion seen at C5/6 and C6/7, causing mild compression over the underlying cord. No cord signal abnormality was seen on imaging. The results of the MRI of the brain showed no notable abnormalities. However, the changes observed on MRI were not considered causative for the lack of sweating (as discussed with the neurologist). Chest X-ray PA view was also done as part of the evaluation and was found to be normal.

A skin biopsy was taken using a 5 mm punch from two different sites on the back for histopathological examination. The first sample was taken from the anhidrotic area on the back, whereas the second was chosen from the normal side. The biopsy from the anhidrotic side showed mild epidermal atrophy with absence of eccrine glands as compared to the normal side. These biopsy findings were atypical for Harlequin syndrome, likely due to sample variability, which may account for the observed histopathological differences between the two sites (Figure 2).

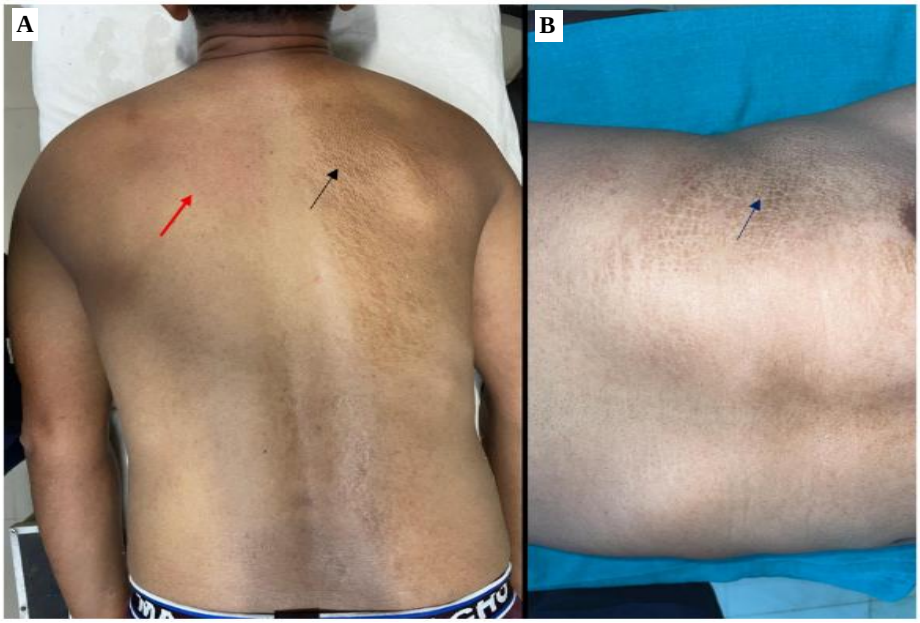


Figure 1: (A) Dry xerotic patches noted on left side of back (black arrow) with mild flushing on left upper back (red arrow); (B) Dry xerotic patches noted on left side of chest (blue arrow).

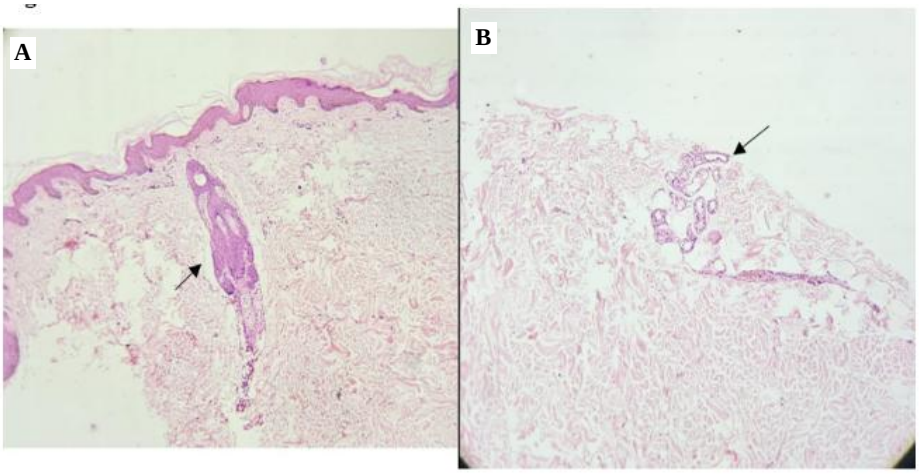


Figure 2: (A) Photomicrograph showing hair follicle with absence of eccrine gland (anhidrotic patch) (H&E stain, 40×). (B) Photomicrograph demonstrating few eccrine glands (normal side) (H&E stain, 10×).

Autonomic function testing, quantitative sudomotor and thermoregulatory sweat testing, and Minor's starch-iodine test were not performed, limiting definitive diagnostic confirmation.

Differential diagnosis includes idiopathic Harlequin syndrome; Ross syndrome; Holmes-Adie syndrome; Horner syndrome; pure autonomic failure; diabetic autonomic neuropathy; Segmental anhidrosis; Syringomyelia; cervical spinal cord lesions; and small fiber neuropathy.

A detailed neurological and ophthalmological examination was performed as part of the comprehensive clinical evaluation to identify any associated neurological or autonomic abnormalities. The examination did not reveal any pupillary abnormalities, abnormal deep tendon reflexes, other neurological deficits, facial sweating abnormalities or evidence of autonomic dysfunction elsewhere. The patient also had no relevant systemic or neurological history that could suggest an underlying secondary cause for the presenting symptoms. MRI evaluation did not demonstrate any structural neurological abnormality or cervical spinal cord lesion that could explain the observed unilateral anhidrosis.

In view of the characteristic unilateral sweating abnormality, absence of neurological or ophthalmological abnormalities, negative relevant history, normal neurological examination and unremarkable MRI findings, the overall clinical assessment was considered consistent with idiopathic Harlequin syndrome after exclusion of clinically relevant secondary causes. The diagnosis was therefore based primarily on the characteristic clinical presentation and the absence of findings suggestive of an identifiable underlying neurological or structural cause.

The patient was counselled regarding the nature and course of the disease. Topical aluminium chlorohydrate lotion was prescribed for the left side of the trunk to manage excessive sweating, while emollients were prescribed for the dry, xerotic patches on the right side. The patient was counselled regarding the benign nature, clinical course, and prognosis of the condition. Appropriate reassurance was provided. However, the patient did not return for subsequent follow-up visits, and long-term clinical outcomes could not be assessed.

DISCUSSION

To date, approximately 100–125 cases of Harlequin syndrome have been documented in the literature, with the majority presenting with facial involvement. The condition predominantly affects females and typically manifests during the third decade of life.[3] Most reported cases are characterized by asymmetric facial flushing and sweating, reflecting the underlying sympathetic dysfunction. In contrast, involvement of other anatomical regions, particularly isolated truncal involvement without associated facial manifestations, is rare and has been infrequently described in the literature. This presentation reflects a unilateral sympathetic dysfunction affecting the normal regulation of sweating. It is characterized by flushing with unilateral hyperhidrosis associated with hypo or anhidrosis and paleness of the contralateral side, due to a compensatory response on the unaffected sympathetically innervated side.[4] The resulting asymmetry in sweating and flushing becomes more evident during conditions that stimulate sympathetic activity. Emotional stress, physical exertion, heat can act as a trigger factors.[5]

Harlequin syndrome is idiopathic in the majority of reported cases, with no identifiable underlying cause despite clinical evaluation. In rare instances, Harlequin syndrome may occur secondary to compressive pathological processes affecting the T2 and T3 sympathetic trunks, including conditions such as medullary astrocytoma, mediastinal schwannoma, apical pulmonary tumors or dissection of the carotid artery.[2] These conditions may interfere with sympathetic nerve pathways, resulting in the characteristic unilateral abnormalities of sweating and flushing. Therefore, careful clinical assessment and appropriate investigations are important to identify or exclude potential secondary causes.

Therefore, chest X-ray PA view and MRI of the cervical spine and head/neck should be performed as part of the diagnostic evaluation to rule out any of the compressive etiologies mentioned above. These investigations may help identify underlying structural abnormalities involving the sympathetic pathways that could contribute to the development of the syndrome. Additional iatrogenic causes have been identified, such as Harlequin syndrome after thoracic sympathectomy, jugular vein catheterization, or anterior mediastinal tumor resection. In addition, neurological conditions such as Guillain-Barré syndrome, diabetic neuropathy or multiple sclerosis can also lead to Harlequin syndrome.[2] It has not been linked to any known genetic abnormalities or hereditary conditions.[6]

For ruling out any secondary association, a thorough clinical examination and all routine investigations should be performed to identify any underlying neurological, structural or systemic abnormalities that may contribute to the condition. The psychosocial impact of Harlequin syndrome, including embarrassment and social discomfort, may be significant enough to warrant therapeutic intervention in some patients, particularly when the symptoms interfere with daily activities and social interactions.[7] The only available treatments for idiopathic cases are contralateral sympathectomy, recurrent stellate ganglion block and botulinum toxin injection, with treatment selection depending on the severity of symptoms and individual patient requirements. In contrast, in cases secondary to a compressive etiology, surgical removal of the compressive cause is the treatment of choice.[8,9] In dermatology, two distinct conditions associated with the term 'Harlequin' include Harlequin

ichthyosis and Harlequin color change. Harlequin Ichthyosis is a rare and severe congenital genetic disorder resulting from mutations in the ABCA12 gene, while Harlequin color change is a benign and self-limiting condition observed in neonates.[10,11]

CONCLUSION

Harlequin syndrome is a rare sympathetic disorder that should be considered in patients presenting with unilateral anhidrosis and flushing. A thorough clinical evaluation, including detailed neurological examination and appropriate investigations, is essential to exclude secondary causes and identify any underlying structural or neurological abnormalities. This case highlights the uncommon presentation of isolated truncal involvement without facial manifestations and emphasizes the importance of clinical recognition of idiopathic Harlequin syndrome, especially in patients with unilateral sweating abnormalities and no identifiable secondary etiology.

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