International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2025: 7.089

Harlequin Syndrome in Guyana: A Case Study and Review

Sheneka A. Ghent[1], Jarrel Watson[2]

[1]MD, MMED (IM/ID), Fellowship in Neurology, Neurology Department, Georgetown Public Hospital Corporation, Guyana
Email: *abigailghent[at]gmail.com*

[2]MD, Consultant Neurologist, Neurology Department, Georgetown Public Hospital Corporation, Guyana
Email: *jarrel22[at]hotmail.com*

Abstract: ***Harlequin syndrome is a rare autonomic disorder caused by disruption of sympathetic pathways supplying the face. This results in one-sided facial flushing and sweating, with opposite-side pallor and dryness. Although usually benign, it may indicate underlying pathology and cause psychosocial distress. We report the case of a 41-year-old Indigenous woman with a six-year history of well-defined episodes of facial flushing and sweating on one side, triggered by heat, activity, or emotional stress. Neurological examination was otherwise normal. Magnetic resonance imaging of the brain and cervical and thoracic spine revealed no abnormalities. In the absence of an identifiable cause, a diagnosis of idiopathic Harlequin syndrome was made. The patient was managed conservatively with reassurance, counseling, and trigger avoidance. This is the first known report of Harlequin syndrome in Guyana. It highlights the need to recognize this rare but usually harmless condition.***

Keywords: Harlequin syndrome, autonomic dysfunction, hemifacial flushing, anhidrosis, dysautonomia, Guyana

1. Introduction

Harlequin syndrome is a rare dysautonomic condition characterized by sharply demarcated hemifacial flushing and hyperhidrosis with contralateral facial pallor and anhidrosis. The disorder results when the sympathetic nerve pathway controlling facial blood vessels and sweating is interrupted. Etiologies include injury, compression, inflammation, or iatrogenic disruption of the T2–T3 sympathetic fibers; yet, up to half of cases have no identifiable cause. The syndrome has also been described following cervical and thoracic surgeries, regional anesthesia, and structural lesions of the brainstem or spinal cord.

Symptoms usually start during exercise, heat, stress, or eating spicy food. Although mostly harmless, this condition can cause distress because of its appearance. Early recognition helps avoid unnecessary worry.

2. Case Presentation

A 41-year-old right-handed Indigenous woman with no known past medical history presented to the neurology clinic with a six-year history of episodic unilateral facial flushing associated with excessive sweating. The episodes showed a sharp midline on her face, often reaching her upper chest, and lasted several hours before stopping on their own.

Symptoms were consistently triggered by strenuous physical activity, exposure to heat, and emotionally stressful situations, particularly anxiety. She reported no headaches, vision changes, weakness, numbness, or other symptoms. She had never smoked and only drank alcohol rarely.

On examination, there was striking hemifacial erythema and hyperhidrosis with a sharp vertical midline demarcation, accompanied by contralateral pallor and anhidrosis. The findings extended to the upper anterior chest. The remainder of the neurological examination, including cranial nerve assessment and motor and sensory testing, was normal.

Magnetic resonance imaging of the brain, cervical spine, and thoracic spine was unremarkable, effectively excluding central or structural causes. A sweat test would have helped confirm the diagnosis, but it was not available. In the absence of an identifiable secondary etiology, a diagnosis of idiopathic Harlequin syndrome was established.

Management focused on patient education and reassurance regarding the benign nature of the condition. Topical glycopyrrolate for symptomatic hyperhidrosis was discussed but was not available locally. The patient received counseling on trigger avoidance and psychosocial support.

3. Discussion

To our knowledge, this is the first documented case in Guyana. Raising awareness is vital in areas with limited resources, as it can help avoid mistakes and unnecessary tests.

The sympathetic pathway supplying the face originates in the hypothalamus, descends to the ciliospinal center of Budge (C8–T2), and exits via the T2–T3 sympathetic fibers before ascending to innervate facial vasomotor and sudomotor structures. Interruption of this pathway leads to ipsilateral facial vasoconstriction and anhidrosis, resulting in pallor and dryness, with contralateral compensatory vasodilation and hyperhidrosis producing flushing and sweating. Extension of involvement may produce symptoms affecting the upper chest or arm if T4 fibers are involved, or Horner syndrome if T1 fibers are affected.

Clinically, Harlequin syndrome is characterized by a sharp vertical demarcation between the two sides of the face, often described as resembling the theatrical “Harlequin” mask. Triggers commonly include heat, exertion, and emotional stress. While idiopathic cases are well documented,

secondary causes must be excluded, as the syndrome may be associated with cervical or thoracic tumors, syringomyelia, cervical disc disease, carotid artery dissection, brainstem lesions, or iatrogenic injury following surgery or regional anesthesia. For that reason, brain and spinal scans are needed before calling a case idiopathic.

Management is largely conservative, focusing on reassurance and education. Symptomatic treatment options include topical anticholinergic agents such as glycopyrrolate, botulinum toxin injections for severe hyperhidrosis, and, rarely, surgical sympathectomy in refractory cases. Psychological support and counseling on trigger avoidance play a critical role in improving quality of life.

4. Conclusion

Harlequin syndrome is an uncommon but distinctive manifestation of autonomic dysfunction. Although typically benign, it may cause significant social and cosmetic distress. Recognizing its features and ruling out other causes are key steps in making the right diagnosis and giving the best care. This report contributes the first documented case of Harlequin syndrome in Guyana and highlights the importance of continued awareness and reporting of rare neurological disorders.

Ethical Statement
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. Institutional guidelines did not require ethical approval for this report.

References

[1] Lance JW, Drummond PD, Gandevia SC, Morris JG. Harlequin syndrome: The sudden onset of unilateral flushing and sweating. *J Neurol Neurosurg Psychiatry.* 1988;51(5):635–642.
[2] Guillot M, Boucau C, André M, Rigoard P. Harlequin syndrome: A rare dysautonomic disorder. *Clin Auton Res.* 2015;25(2):115–118.
[3] Naver HK, Sander B, Petersen P, Fuglsang-Frederiksen A. Harlequin syndrome: A clinical, physiological and autonomic study of 6 cases. *J Neurol.* 1990;237(7):410–413.
[4] Lo CP, Chen CY, Lee KW, Hsu WY. Harlequin syndrome associated with cervical sympathetic chain schwannoma. *AJNR Am J Neuroradiol.* 2006;27(6):1308–1310.
[5] Tannoury E, Chen S, Herial NA, et al. Harlequin syndrome after cervical epidural catheter placement: Case report and review of the literature. *Reg Anesth Pain Med.* 2000;25(3):313–316.
[6] Coorey G, Ng AC, Nandurkar D. Idiopathic Harlequin syndrome: Clinical features and management. *Intern Med J.* 2012;42(3):341–343.
[7] Thompson HS, Kardon RH. Horner's syndrome. In: Miller NR, Newman NJ, eds. *Walsh and Hoyt's Clinical Neuro-Ophthalmology*. 6th ed. Philadelphia: Lippincott Williams & Wilkins; 2005. p. 439–495.