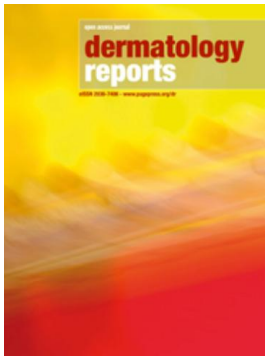

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Unilateral facial flushing and anhidrosis: a case of idiopathic Harlequin syndrome with successful botulinum toxin treatment

Pasquale Sorrentino,[1] Gaetano Licata,[2] Maria Maddalena Nicoletti,[1] Margherita Malaspina,[1] Giuseppe Argenziano,[1] Eugenia Veronica Di Brizzi,[1] Caterina Mariarosaria Giorgio[1]

Dermatology Unit, University of Campania "Luigi Vanvitelli", Naples; [2]Dermatology Unit, San Antonio Abate Hospital, Trapani, Italy

Correspondence: Pasquale Sorrentino, University of Campania "Luigi Vanvitelli", Via Pansini 5, 80131, Naples, Italy. E-mail: passor998@gmail.com

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Consent for publication: the patient gave her written consent to use her personal data for the publication of this case report and any accompanying images.

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Dear Editor,

Harlequin syndrome is a rare autonomic disorder characterized by unilateral facial flushing and sweating caused by dysfunction of sympathetic innervation. Although generally considered a benign condition, it may occasionally represent the first manifestation of underlying neurological or structural abnormalities, including brainstem lesions, multiple sclerosis, or thoracic spinal cord disorders. Due to its striking clinical appearance, Harlequin syndrome may be misinterpreted as an allergic reaction, chronic urticaria, or other dermatological conditions, potentially delaying diagnosis. We report a case of idiopathic Harlequin syndrome in a middle-aged woman, highlighting the importance of recognizing this uncommon autonomic disorder in dermatological practice.

A 57-year-old woman with no relevant dermatological history presented with a 6-month history of recurrent episodes of unilateral facial flushing and sweating. Episodes were consistently triggered by physical exertion or emotional stress and resolved spontaneously. The patient denied previous similar manifestations, allergies, medication use, or exposure to potential triggering substances. Her medical history was notable for cluster headaches occasionally associated with lacrimation and nasal discharge, but no additional neurological symptoms were reported (Figure 1).

Dermatological examination revealed unilateral erythema and sweating involving the right hemiface, whereas the contralateral side appeared pale and dry. Neurological examination showed no focal deficits, and autonomic reflexes were preserved. Based on the asymmetric vasomotor and sudomotor response, differential diagnoses included allergic reactions, chronic inducible urticaria, angioedema, and neurological disorders. The absence of pruritus, wheals, mucosal involvement, or systemic symptoms made allergic and inflammatory conditions unlikely. Furthermore, the reproducible association with physical activity and emotional stimuli, together with the presence of contralateral anhidrosis, suggested an autonomic dysfunction rather than a primary dermatological disorder.

A comprehensive diagnostic workup was performed to identify possible secondary causes. Contrast-enhanced computed tomography of the mediastinum excluded masses or vascular abnormalities potentially compressing the sympathetic pathway. Magnetic resonance imaging of the brain, cervical spine, and thoracic spine with contrast revealed no evidence of demyelinating disease, syringomyelia, or structural lesions. A quantitative sweat test confirmed left-sided anhidrosis, consistent with sympathetic dysfunction. After neurosurgical consultation excluded a compressive etiology, a diagnosis of idiopathic Harlequin syndrome was established.

Given the relevant impact of symptoms on the patient's quality of life, treatment with botulinum toxin injections was performed in the affected hemifacial area, resulting in a marked reduction of both flushing and sweating, with significant clinical improvement (Figure 1).

Harlequin syndrome results from interruption of sympathetic pathways extending from the hypothalamus through the brainstem and spinal cord to the cervical sympathetic chain and facial structures. Depending on the anatomical level involved, lesions may occur in the brainstem, spinal cord, thoracic sympathetic roots (T1–T3), cervical sympathetic chain, or along the carotid artery. In our patient, extensive imaging excluded structural abnormalities, supporting the diagnosis of an idiopathic form.

Nevertheless, long-term clinical follow-up remains important, as Harlequin syndrome may occasionally precede or accompany neurological diseases, including Guillain-Barré syndrome, diabetic autonomic neuropathy, and Bradbury-Eggleston syndrome. In addition, recent reports have expanded the spectrum of associated conditions, describing Harlequin syndrome in critically ill patients with autonomic instability, including those receiving venoarterial extracorporeal membrane oxygenation.[1] Post-surgical cases involving thoracic procedures have also been reported, suggesting that sympathetic pathway injury may be an underrecognized mechanism in selected clinical settings.[2]

Management of Harlequin syndrome is primarily symptomatic and should be tailored according to disease severity and patient expectations. Pharmacological treatments, including beta-blockers such as propranolol, may reduce excessive flushing, while anticholinergic agents such as oxybutynin can decrease sweating. Among minimally invasive approaches, botulinum toxin represents a valuable therapeutic option due to its ability to inhibit acetylcholine release and reduce eccrine gland activity. In our patient, botulinum toxin injections provided sustained improvement, allowing better control of symptoms and improving quality of life.

This case emphasizes the importance of considering Harlequin syndrome in patients presenting with unilateral facial erythema and sweating. Although idiopathic forms usually have a favorable prognosis, appropriate neurological evaluation and imaging are essential to exclude potentially serious secondary causes. Awareness of this rare entity among dermatologists may facilitate prompt recognition, adequate investigation, and effective symptomatic treatment.

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Figure 1. A clinical image of Harlequin syndrome: a 57-year-old woman presents with post-exercise hot flashes and sweating limited to the right half of her face; the pupils are normal.