

Harlequin Syndrome with Crossed Sympathetic Deficit of the Face and Arm

Harlequin syndrome is characterized by unilateral hyperhidrosis and flushing, which are predominantly induced by heat or exercise. Usually, the sympathetic deficits confine to the face. Rarely, the autonomic deficits involve the arm or the parasympathetic neurons in the ciliary ganglia. We report a 43-yr-old woman who presented with facial flushing and sweating in the right side, which were mainly induced by exercise. The facial flushing accompanied relative coldness in the right arm. Valsalva maneuver, cold pressure and 0.125% pilocarpine test, and computed tomography of the chest were normal. The crossed sympathetic deficit in the left face and right arm suggested that the lesions were multifocal. The sympathetic impairment in our patient may lie on a spectrum of pre- and postganglionic autonomic dysfunction, which was observed in Holmes-Adie, Ross, and Guillain-Barré syndrome.

Key Words : Harlequin Syndrome; Hypohidrosis, Autonomic Neuropathy; Hereditary Sensory and Autonomic Neuropathies

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INTRODUCTION

Harlequin syndrome is characterized by unilateral facial flushing and sweating, which are predominantly induced by heat or exercise (1). Harlequin syndrome is mostly idiopathic (1), but may occur in association with brainstem infarction, superior mediastinal neurinoma, internal jugular vein catheterization (1-3). The sympathetic deficit is in the non-flushing side and the healthy side shows normal or excessive flushing and sweating. Usually the sympathetic deficit is confined to the face. Rarely, the autonomic deficit involves the arm or the parasympathetic neurons in the ciliary ganglia (4, 5). We report a patient with unilateral facial flushing and sweating, and ipsilateral arm coldness, which were mainly induced by exertion.

CASE REPORT

A 43-yr-old woman presented with a 3-yr-history of facial flushing and sweating in the right side, which were mainly induced by exercise. She had also noticed an increased coldness of the right arm during such episodes. The symptoms were more prominent in the winter, but did not aggravate during the preceding three years. Her family and medical history were unremarkable. Findings of the physical and neuro-

logical examinations at rest were normal. However, after a 30-min-running, she showed flushing and sweating only on the right side of the face (Fig. 1). During this episode, the left side of her face remained dry with impaired sweating. Pupils were equal. Blood pressure, pulse rate, and skin color did not differ among the four extremities during the episode. Using a digital skin thermometer, skin temperature measured 2°C colder on the right arm than on the left arm, and 2.9°C colder on the left face than on the right face. Response to the Valsalva maneuver and sympathetic skin response were normal. The immersion of one hand in ice water for 1 min (cold pressure test) revealed a normal response in both arms. The pupils did not constrict with 0.125% pilocarpine eyedrops. Computed tomography of the chest was normal.

DISCUSSION

The name harlequin syndrome was first coined by Lance and Drummond in 1988 when they described five cases of unilateral facial flushing and sweating, which were induced by exercise in four (1). They proposed that the torsional occlusion of the anterior radicular artery at the third thoracic segment caused the syndrome, and that the lesion might involve both pre- or postganglionic cervical sympathetic fibers and parasympathetic neurons of the ciliary ganglion.



Fig. 1. Photograph of the patient, taken after a 30-min-run, shows facial flushing only in the right side. During this episode, the left side of her face remained dry without sweating.

The preganglionic neurons of the sympathetic division originate in the intermediolateral cell column of the spinal gray matter, from the eighth cervical to the second lumbar segments. The preganglionic fibers synapse with the cell bodies of postganglionic neurons, which are collected in two large ganglionated chains or cords, one on each side of the vertebral column (paravertebral ganglia), and in several single prevertebral ganglia. There are three cervical (superior, middle, and inferior or stellate), eleven thoracic, and four to six lumbar sympathetic ganglia. The head receives its sympathetic innervation from the eighth cervical and the first two thoracic cord segments, the fibers of which passing through the inferior and middle cervical ganglia, and synapsing at the superior cervical ganglia. Postganglionic fibers from the cells of the superior cervical ganglion follow the internal and external carotid arteries and innervate the blood vessels and smooth muscle as well as the sweat, lacrimal, and salivary glands of the head. The arm receives its postganglionic innervation from the inferior cervical and uppermost thoracic ganglia, which are fused to form the stellate ganglion. Our patient showed crossed sympathetic deficits in the left face and right arm, which suggested that the lesions were multifocal. Previously, crossed sympathetic deficits involving the face, arm, and leg has been described only once, in a 37-yr-old woman (6). Caparros-Lefebvre et al. (7) reported another patient with unilateral loss of facial flushing and sweating, and with contralateral anhidrosis, which were induced by exercise. However, she also had tonic pupils and areflexia, and was consid-

ered to have Ross syndrome (Adie syndrome with anhidrosis).

The spectrum of pre- and postganglionic autonomic dysfunction in patients with harlequin syndrome indicates a relation with Holmes-Adie and Ross syndromes, and the persisting autonomic deficit in Guillain-Barré syndrome (8-10). The clinical course of partial dysautonomias, such as Ross syndrome or Holmes-Adie syndrome, seems to be benign but may progress slowly. Each of these entities has a more widespread dysfunction than previously believed, with considerable overlap. There is increasing evidence that classic partial autonomic disorders may lie on a spectrum of generalized injury to the autonomic and dorsal root ganglia (11, 12). Because the sympathetic, parasympathetic, and dorsal root ganglia are all neural crest cell derivatives, it is conceivable that some property of these related tissues makes them more susceptible to injury, although the mechanism of ganglionic damage or degeneration remains unknown.

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