A Case of Harlequin Syndrome

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= ABSTRACT =

Asymmetrical facial sweating and flushing induced by heat, stress and exercise have been named as Harlequin syndrome. The Harlequin syndrome represents a local autonomic dysfunction due to a pre or postganglionic cervical sympathetic deficit on the non-flushing side. It is unusually caused by the structural lesions detected by CT or MRI. We here report a case of well-demarcated, right hemifacial flushing and sweating following chiropractic maneuver for intercostal neuralgia.

(Key Words: Harlequin syndrome, Chiropractic)

Introduction

Harlequin syndrome is caused by an interruption of ipsilateral sympathetic supply to the face, resulting an abnormal pale and dry skin on the affected side with a normally flushed and sweaty appearance on the unaffected side. To date, only several cases have been reported, suggesting it is a rare disease or its incidence has been underestimated.

The sympathetic nervous system mediates thermally induced facial sweating and flushing, and the hemifacial loss of sweating and flushing is commonly accompanied by the signs of ocular sympathetic deficit (Horner’s syndrome). However it has been also reported in association with tonic pupils (Ross’s syndrome) or with apparently normal ocular sympathetic innervation (Harlequin syndrome). Here, we reported a case of Harlequin syndrome and reviewed the literatures.

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Case

A 40 year-old woman had been in good health until 3 years before when she began to have right hemifacial flushing and sweating induced by exercise and hot bath. Previously, she had been suffering from right upper thoracic intercostal neuralgia in her thirties, which initially had been treated with gabapentin for 6 months without benefit. Then, she began to take a thoracic spinal chiropractic treatment for several times and her pain improved gradually. The next day she began to notice a sweating on her left face and difference in skin colors between each side of face, which persist after strenuous exercise and hot bath.

A general physical examination was normal. On neurological examination, pupils were equal in size, round and reacted to light without ptosis. We investigated pupillary reactivity by phenylephrine and pilocarpine eye drops, which revealed no evidence of a sympathetic or parasympathetic innervation abnormality, including Horner's syndrome. The remainders of the cranial nerve examination were normal, and there was no motor or sensory deficit. Deep tendon reflexes were normal (2+) throughout. Plantar responses were flexor. Coordination and gait were normal. A photograph (Fig. 1) was taken after 20-min run. It showed a distinct line of demarcation between halves of her face, showing red and sweating on the right and pale and dry on the left. No other symptoms were accompanied. There was no relevant injury or histories of intercostal nerve block or sympathetic block. Brain and cervico-thoracic spinal magnetic resonance imaging (MRI), somatosensory evoked potentials, and cardiovascular autonomic reflexes were all normal. CSF study including PCR for herpes virus, PRP and routine laboratory was normal. Her blood pressure was 120/80mmHg.

Discussion

Hemifacial loss of sweating and flushing with normal ocular sympathetic innervation called Harlequin syndrome. Harlequin syndrome in adults was first described by Lance et al.\textsuperscript{1} as the sudden onset of unilateral facial flushing and sweating.

There are several autonomic syndromes associated with unilateral facial flushing. Along with the distribution of flushing, and the presence or absence of associated autonomic signs, the primary lesion can be localized along a three-neuron pathway in different syndromes. Central sympathetic lesions may occur with brainstem infarction or with migraine, producing a central Horner's syndrome or a Harlequin syndrome accompanied by hemi-body loss of thermoregulatory sweating\textsuperscript{1,8}. The second order preganglionic pupillomotor fibers leave the spinal cord in the first thoracic root, whereas sudomotor and vasomotor fibers leave below T1, at T2 and T3. Using the techniques of stimulation and ablation of the exposed