

Harlequin Syndrome: not just a Color Change

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Abstract

Harlequin syndrome is an autonomic disorder of the sympathetic vasomotor and sudomotor nerve fibers characterized by unilateral facial flushing and hyperhidrosis with contralateral anhidrosis. These symptoms are induced by heat, physical activity or emotional factors. Trunk and arms can also be affected. We report a case of a 46-yr-old woman who was referred to UHC, Dermatology Department with complaints of left side facial erythematous flushing and sweating during physical activities especially in hot weather and the right side of the face remaining dry and maintaining its normal color. Detailed examinations were made and no structural abnormality or any other obvious cause of the condition was identified. After the exclusion of

other diagnoses, we concluded the diagnosis Harlequin Syndrome.

Practice Points

- Harlequin Syndrome consist on a unilateral facial flushing and sweating triggered by exercise or hot weather
- CT scan or MRI are important for the differential diagnosis.
- Mostly no treatment is recommended.

Key words: Harlequin syndrome, erythematous flushing, unilateral sweating.

INTRODUCTION

Harlequin syndrome was first described by Lance et al. in 1988 as the sudden appearance of erythematous flushing and sweating limited only in one side of the face in response to heat or exercise. It was named Harlequin syndrome based on the character from the Italian theatre “Commedia dell’ Arte”. It is mostly an idiopathic condition although organic causes must be excluded. Lance et al. proposed the occlusion of the anterior radicular artery during strenuous exertion as the pathological mechanism (1). Harlequin syndrome may also be associated with Horner syndrome, Adie syndrome and Ross syndrome and it is named as Harlequin sign (2).

CASE REPORT

A 46-yr-old woman presented in the UHC “Mother Theresa” Dermatology Department with one year history of unilateral facial flushing and sweating affecting only the left side of the face. At the same time the other side remaining pale and dry. These symptoms were provoked by physical exercise during hot weather or emotional stress. Her previous medical history revealed a hospitalization in pulmonary hospital before she presented to our department. Patient had been admitted to the Department of Pneumology for exertion dyspnea and was diagnosed and treated for First Stage Sarcoidosis. Thoracic CT scan had revealed only hilar and paratracheal lymph nodes enlargement. She had no other significant medical history. There was no history of undergoing any surgical procedure. Her family medical history

was unremarkable. On physical examination no sweating or flushing was noted at rest. Her vital signs were normal. No pupillary abnormalities including signs of Horner syndrome were observed. A detailed eye examination performed by an ophthalmologist revealed no abnormalities. Laboratory evaluation including complete blood count, ESR, blood sugar, renal, hepatic and thyroid function tests were all normal. ANA was found to be positive (++) . During the neurological examination, including cranial nerves exam, motor and sensory systems evaluation, higher mental function, deep tendon reflexes did not result any abnormality. The symptoms (an erythematous flushing and sweating on the left side of her face) were evident after a 30-minutes excessive walking or running in hot weather. These symptoms were alleviated by cold compresses and after stopping the physical activity. During the episode the pupils were equal in size and reacted normally to light. Her vital signs were also normal. A CT scan of the head and neck revealed only bilaterally latero-cervical lymph node enlargement of 6 mm without any structural lesion. A Doppler ultrasound of the carotid arteries resulted normal. The diagnosis of Harlequin syndrome was made by excluding other possible diagnosis. The patient was reassured of the benign nature of this condition. A possible solution such as sympathectomy lateral to the flushing side was suggested to the patient to help relieve the symptoms.



Figure 1. Evident hemifacial erythema just in one side

COMMENTS

In this short paper we present a case as a classic manifestation of Harlequin syndrome. Asymmetrical facial sweating and flushing has been named the "Harlequin Sign" (1, 3). Harlequin syndrome is a rare autonomic disorder first described by Lance et Drummond 1988 as a manifestation of unilateral facial flushing and sweating during hot weather or when exercising (1). It was first thought that the occlusion of an anterior radicular artery during exercise was the pathogenic mechanism to explain the symptoms, but the real origin of this syndrome is a dysfunction of the sympathetic chain. The neural

damage of the sympathetic system affects the non-flushing side and the flushing side of the face (arms and trunk sometimes) is known to be a compensatory over reaction (4).

Is Harlequin sign just a color change? When an acute unilateral color change occurs in a patient's appearance, it should be first considered a neurovascular cause. As such a cause may be immediately life threatening, neurovascular disease should be excluded before other potential causes are considered. According to case reports, unilateral facial flushing can be a sign of acute stroke, possibly due to failure of the autonomic nervous system (5).

Different diagnosis taken in consideration are: Horner syndrome with miosis, ptosis and enophthalmos which may indicate a lesion of the superior cervical ganglion; Ross syndrome with tonic pupils, anhidrosis and hyporeflexia which may indicate a lesion of the postganglionic cholinergic parasympathetic and sympathetic fibers projecting to the iris (2). Referring our case Ross syndrome and Horner syndrome were excluded by the ophthalmological and neurological examination performed that showed no such abnormalities.

Idiopathic etiology is frequent in older children and adults, but other possible causes including iatrogenic lesions and malignances should be excluded. There are case reports of Harlequin syndrome due to epidural anesthesia or other procedures in the cervicothoracic region (5). More alarming conditions such as neoplasm of superior mediastine and apical lung can be behind

a Harlequin sign (6, 7). Other less frequent causes are neurotropic virus infection, autoimmune diseases as multiple sclerosis, syringomyelia (2, 7).

What is important to do in a case with Harlequin sign? The diagnostic process is important for ruling out malignant causes or iatrogenic ones. A thorough physical, neurological, and ophthalmological examination is essential and followed by radiology tests (7).

In our patient the thoracic and cranio-cervical CT scan performed showed no space occupying lesion. The cervical lymph nodes noticed to the CT scan were due to sarcoidosis diagnosed before. Except for examination ANA positivity no other abnormality was found. Reviewing the literature we found many cases of autoimmune origin for such syndromes (8, 9, 10) and concluded of a possible autoimmune origin for the symptoms of our patient. She did not accept to receive any treatment. We recommended to follow up.

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