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Case Report

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Harlequin Syndrome Secondary to Herpes Simplex Encephalitis

Abhishek Dixit[1], Abhishek Pathak[1], Anand Kumar[1], Rameshwar Nath Chaurasia[1], Vijaya Nath Mishra[1], Deepika Joshi[1], Varun Kumar Singh[1*] and Pratishtha Sengar[2]

[1]Department of Neurology, Institute of Medical Science, Banaras Hindu University, India
[2]Department of Pathology, Heritage Hospital, India

Abstract

Background: Harlequin syndrome is a rare dysautonomia syndrome characterized by increased sweating on one side and anhidrosis on the contralateral side of the body.

Case report: A 24 years old male presented with fever, multiple episodes of focal seizures with bilateral tonic clonic movements and altered sensorium. On evaluation, he was diagnosed as a case of Herpes Simplex Encephalitis based on classical radiological and positive cerebrospinal fluid real time polymerase chain reaction test for herpes simplex virus 1. He received full course of intravenous acyclovir during hospital stay and discharged on two antiepileptics. During follow up, he developed increased sweating on one half of face and anhidrosis over another half suggestive of Harlequin syndrome. However, these symptoms subsided by its own without any active intervention during further follow up.

Conclusion: To the best of our knowledge, this is the first report of Harlequin syndrome secondary to herpes simplex encephalitis.

Keywords: Harlequin Syndrome • Herpes Simplex Encephalitis • Dysautonomia • Sweating

Introduction

Harlequin syndrome, a rare dysautonomia syndrome of the face, characterized by increase in sweating of one side and anhidrosis of the opposite side of the body [1]. It often affects upper thoracic region of the chest, neck and face. The efferent sympathetic sudomotor signal originates in the cerebral cortex. It passes through the hypothalamus, brainstem and spinal cord and exits to synapse in the sympathetic chain. These signals travel through post-ganglionic fibers and reach the sweat glands in the skin [2]. The chief controller of this thermoregulatory response is the preoptic hypothalamic area [3]. Harlequin syndrome occurs by interruptions in this pathway which results in reduction in response to thermal or psychogenic stimuli. We report the first case of Harlequin syndrome secondary to Herpes Simplex Encephalitis (HSE).

Case Report

A 24 years old right handed male presented with acute onset, moderate grade fever for 8 days, multiple episodes of focal seizures with bilateral tonic clonic movements and altered sensorium for 4 days. There was no history of cough with expectoration, skin rash, and myalgia, bleeding manifestations, joint pain or ear discharge. There was also no previous history of tubercular contact, jaundice or chronic drug intake. Patient had no previous co-morbidities. Family history was negative for similar illness. He was receiving two antiepileptics, injection levetiracetam 1000 mg/day and injection lacosamide 200 mg/day from outside. On examination, Glasgow Coma Scale (GCS) was E4V1M6. Pulse rate was 78/min, axillary temperature 39°C, blood pressure 122/70 mmHg, and respiratory rate 20 min with normal oxygen saturation. There was no ptosis. Pupils were bilaterally symmetrical, normal in size and reaction to light. Papilledema was absent. Muscle power, tone and deep tendon reflexes were normal in all four limbs. Cardiovascular, respiratory and abdominal examinations were within normal limits.

His complete blood count, blood sugar levels, renal and liver function tests, chest radiography and electrocardiography testing were within normal limits. His blood and urine culture, dengue and malaria serology were negative. Cerebrospinal fluid (CSF) analysis revealed protein-104.7 mg/dl, cell count-27(100% lymphocyte) and glucose 47 mg/dl. Magnetic resonance imaging (MRI) brain revealed T2W/FLAIR hyperintensities in left temporal lobe, left orbitofrontal lobe and left insular cortex with gyral swelling in right temporal lobe (Figure 1). Real time polymerase chain reaction (PCR) testing of CSF was positive for Herpes Simplex Virus 1. A final diagnosis of HSE was made. Intravenous acyclovir 500 mg three times a day was given for 21 days. Injection levetiracetam and lacosamide were continued. On day 18 of admission, his GCS improved to E4V5M6. No further episodes of fever and seizures occurred. On higher mental function testing at discharge (day 23), lexical fluency, calculation, registration and recall were impaired.

At the end of two weeks follow up, relative noticed increased sweating over right side of face and neck of patient. It increased with emotional stress and on sunlight exposure. There was no sweating on the left side. He reported no other local or systemic symptoms (Figure 2). These symptoms were consistent with the diagnosis of Harlequin syndrome secondary to herpes simplex encephalitis. At 3 months of follow up, these symptoms subsided by its own.

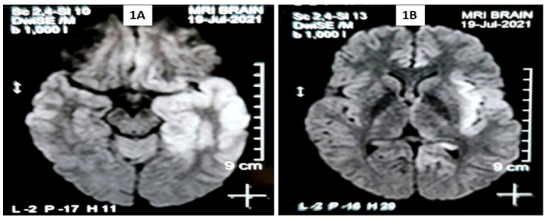

Figure 1. Magnetic resonance imaging of brain showed hyperintensities in

***Address for Correspondence:** Varun Kumar Singh, Department of Neurology, Institute of Medical Science, Banaras Hindu University, India, Email: mailurvarun@gmail.com*

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left temporal lobe, left orbitofrontal lobe and left insular cortex (1A and 1B) with associated gyral swelling in right temporal lobe (1A).

Figure 2. Patient demonstrating sweating only on the right side of the face (2A) with sparing of left side (2B).

Results and Discussion

The above case presented with acute onset fever, focal with bilateral tonic clonic seizures, and altered sensorium. A diagnosis of HSE was made based on typical radiological findings and positive CSF PCR for HSV1. In follow up, he was diagnosed as a case of Harlequin Syndrome.

"Harlequin syndrome" was first reported by Lance et al in 1988 in a patient of autonomic disturbance due to hemifacial cutaneous denervation [4,5], which led to unilateral anhidrosis and reduced or absent facial flushing on affected side of the face [5]. Although the preoptic hypothalamic area plays an important role in the regulation of sweating, a lesion in any area of the sympathetic sudomotor pathway can cause an abnormal sweat response. In the brain, the amygdala, hippocampus, and cingulate gyrus are involved in sweat response [6].

Lentiform nuclei and anterior hypothalamus/preoptic area are primary responsible for the thermogenic sweating and amygdala and hippocampus for psychogenic sweating. How these brain regions influence the sweating regulation is still unclear [7]. Most of the Harlequin syndrome cases are primary in nature. Structural pathology (secondary harlequin syndrome) causing damage to sympathetic outflow are reported in about one sixth of the patients only [8]. The iatrogenic type secondary to surgical procedures has been most frequently reported [9]. The primary sites involved in Herpes Simplex Encephalitis are the limbic system, medial temporal lobes, insular cortices, and inferolateral frontal lobes [10]. Some of these regions are also related to sweat regulation. Yang TW et al reported hemifacial anhidrosis in a case of herpes simplex encephalitis having typical radiological finding in right medial temporal lobe including amygdala and hippocampus, which improved with acyclovir [11]. Hence, involvement of bilateral temporal lobes and left insular lobe in the present case of herpes simplex encephalitis might explain harlequin syndrome manifestation.

Majority of these patients do not need medical or surgical interventions, but only treatment of the underlying condition. For cosmetic indication, patient needs psychological support and may be considered for contralateral sympathectomy.

Conclusion

To the best of our knowledge, this is the first case report of Harlequin syndrome secondary to Herpes Simplex Encephalitis.

Conflicts of Interest

No potential conflict of interest is relevant to this article.

Funding Support

No specific funding was received for this work.

Ethics Statement

Ethical approval for this study was not needed as per institutional ethics policy, as this study is a case report of a single patient and did not include protected health information, data analysis, or testing of a hypothesis. Written informed consent from the patient for the study and the publication was obtained.

Author's Contribution

Varun Kumar Singh: Concept and study design, manuscript writing

Abhishek dixit: Data collection, manuscript writing

Abhishek Pathak: Manuscript writing and editing

Anand Kumar: Manuscript writing and data collection

Rameshwar Nath Chaurasia: Manuscript writing and editing

Vijaya Nath Mishra: Manuscript writing and data collection

Deepika Joshi: Manuscript writing and editing

Pratishtha Sengar: Manuscript writing and data collection

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