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Case Report

Continuous Unilateral Cephalalgia Due to Systemic Lupus Erythematosus: A Case Report

Lucas Teixeira, Matheus Cruz

Universidade Federal do Rio Grande do Sul, Brazil

Lucas Teixeira
lfteixeira@hcpa.edu.br

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Marcelo Moraes Valença

Abstract

Background
Hemicrania continua is a rare form of cephalalgia featuring a chronic and persistent headache in only one side of the head.

Objectives
In this report, we present a case of a patient with hemicrania continua and systemic lupus erythematosus (SLE).

Methods
We collected patient data through the electronic medical record. Afterward, we reviewed the literature regarding hemicrania continua and its pathophysiology and correlation with neurovascular alterations, inflammation, and SLE.

Results
A 42-year-old woman visited the emergency department due to worsening constant unilateral cephalalgia that had been present for the past 6 months. The patient reported a highly intense (10/10) headache in the entire left hemicrania that radiated to the left shoulder. During physical examination, she presented with nystagmus, vertigo, and aggravated cephalalgia associated to body movement and, despite having no optic nerve thickening. In addition, she had jaundice, tachycardia, and splenomegaly. Complimentary exams found deep anemia, depletion in complement system and anti-nuclear factors, suggesting a possible hemolytic anemia (AIHA) due to SLE. Treatment was initiated with hydrocortisone and prednisone, associated with amitriptyline, fluoxetine and diazepam, reaching full remission.

Conclusion
These syndromes have aggravated each other, and possibly the explanation for the cephalalgia remission was the control of AIHA and SLE. It features a rare case in literature and thus warrants discussion.

Keywords:
Headache
Unilateral cephalalgia
Systemic lupus erythematosus
Corticosteroid therapy
Inflammation

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Introduction

Hemicrania continua is a rare form of cephalalgia featuring a chronic and persistent headache in only one side of the head. There are associated symptoms such as nausea and vomiting, throbbing pain, ptosis, miosis, and nasal congestion. It is difficult to get to a clinical diagnosis since most clinicians are not aware of hemicrania. Therefore, further data on this disease is necessary to describe the clinical features of it. The pathophysiology of hemicrania has not yet been fully understood. Neurovascular alterations have been described as a possible cause for primary headaches, but the understanding of the mechanism is still unknown. This correlation between primary headaches and neurovascular alterations has already been described (1). Inflammation may cause headache itself or even(2)or even be a trigger for neurovascular alterations and thus headaches. This discussion warrants further research on the theme.

Case report

A 42-year-old woman visited the emergency department due to worsening constant unilateral cephalalgia that had been present for the past 6 months. The patient reported a highly intense (10/10) headache in the entire left hemicrania that radiated to the left shoulder and did not alleviate with analgesic pills. In the last 2 months, she experienced extreme nausea, significant weight loss, blurred vision in the left eye, vertigo, pain worsening after movement, and episodes of self-mutilation to distract from the severe headache. She had an irregular menstrual cycle, had undergone 2 abortions, and had no children. During physical examination, she presented with nystagmus, vertigo, and steep cephalalgia when assessing eye movement, despite having no optic nerve thickening. In addition, she had jaundice, tachycardia, splenomegaly, Traube's space dullness on percussion, and significant abdominal pain in both upper quadrants after deep palpation.

This complex case involves an intersection of more than two syndromes. The patient fits the criteria for hemicrania continua according International Classification Headache Disorders (ICDH-3), since she had persistent unilateral headache for more than three months associated with a sense of aggravation of the pain by movement (3, 4). She also would fit the criteria for systemic lupus erythematosus (SLE), according the European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification, since the patient had nonscarring alopecia, autoimmune hemolysis, low C3 and C4, and high titer of Anti-Nuclear Antibodies (ANA) (5). Complimentary exams revealed that the patient had autoimmune hemolytic anemia (AIHA), as evidenced by low hemoglobin level (4.2 g/dL; normal range >11.6 g/dL) and elevated indirect bilirubin level (2 mg/dL; normal range <0.7 mg/dL). Her AIHA may have been triggered by SLE, which was diagnosed based on her low C3 and C4 levels (77 and 10.3 mg/dL, respectively; normal ranges 82-193 mg/dL and 15-57 mg/dL, respectively) and high ANA titer (1/320 nuclear AC-4 and AC-7 pattern). Other clinical features of SLE included alopecia, splenomegaly, weight loss, and a history of miscarriages. However, the continuous unilateral cephalalgia had no defined cause since it had no previous trigger event or image exam alteration, being linked to a possible relation to AIHA subsequent to SLE disruption. Infectious diseases, thrombosis and aseptic encephalitis were rejected as major causes after thorough serology testing and image exams. SLE is an autoimmune disease that can cause vascular inflammation and damage to multiple organs, including the spleen, liver, and serosa (the lining of the pleural and pericardial cavities). Another presentation is neurologic lupus, which is related to psychosis and has been described in cephalalgia crisis. This inflammatory and vasculitis component may explain the intense unilateral headache described in this case since SLE is a condition strongly related to inflammation, and so is cefalalgia (6-8). The mechanism of SLE cephalalgia is not fully understood. Still, it may be related to impaired neuronal excitability, which is a major component of cephalalgia, and inflammation, which has been associated with chronic migraine's pathophysiology. The neurons in SLE are damaged due to inflammation, blood-brain-barrier lesions and release of cytokines such as tumor necrosis factor-α. A treatment for SLE would be an interesting way to mitigate inflammation that worsen chronic migraine (9-11). After treatment of AIHA and SLE with hydrocortisone 100 mg daily, then prednisone 60 mg twice a day, the patient was started on daily amitriptyline 5 mg, diazepam 5 mg, and fluoxetine 20 mg, which resulted in complete symptom remission. While corticosteroids improved autoimmune damage caused by SLE, the psychodrugs affected the unilateral headache. This dual front management is a reliable asset when it is necessary to control an intense inflammatory component. It is a relatively simple way to manage a complex situation and cause clinical improvement. The patient referred no more complaints two weeks after the treatment began and has been stable since. Although SLE patients may describe cephalalgia and chronic migraine, this case report of continuous unilateral cephalalgia in SLE is rare and warrants further study.

Discussion

This case report describes a patient with an unusual clinical presentation of an inflammatory disease. The patient's cephalalgia was probably aggravated by her onset of SLE, and the explanation for the chronic unilateral cephalalgia remission may be the control of AIHA and SLE using corticosteroids, along with

benzodiazepines and antidepressant combination. SLE inflammatory response was controlled, as evidenced by the reduction of inflammatory markers, and during the same period, the patient stopped reporting intense cephalalgia, suggesting that the improvement of one condition caused remission of the other one. Although SLE has been extensively related to psychosis and other neurologic symptoms, a correlation to chronic unilateral cephalalgia has not been described. It features as a rare case in literature and thus warrants discussion.

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