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A State of Possession-Like in a Patient with Flare of Systemic Lupus Eritematosus

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Abstract

Introduction: Symptoms and signs related to a state of “possession” have been associated with psychological disorders, but have not been described as manifestations of organic cerebral disease. Method: Case report of a 17 year-old female patient with episodic symptoms of a possession-like state manifested as aggressiveness, tremor, upper eye deviation, incoherent language, aversion to religious symbols, and demanding to be called by other names. Results: These symptoms were present during episodes of orthostatic hypotension, as a result of autonomic neuropathy developed in the context of a flare of systemic lupus erythematous (SLE). Single photon emission computerized tomography (SPECT) with 99mTc-HmPAO in upright position showed an extensive ischemic area in the left brain. This study was normal in horizontal position, with normal blood pressure and normal mental status. Conclusion: An organic, cerebral-vascular cause of a possession-like state is described.

Key words: Conduct disorder, cerebrovascular circulation, systemic lupus erythematous, orthostatic hypotension.

Título: Un estado similar al de una “posesión” en una paciente con recaída de lupus eritematoso sistémico

Resumen

Introducción: Los síntomas y signos relacionados con un estado de “posesión” se han asociado a trastornos psicológicos, pero no se han descrito como manifestaciones de una afección orgánica cerebral. Método: Reporte de caso de una mujer de 17 años de edad con síntomas
episódicos similares al de un estado de la posesión, manifestado por agresividad, temblor, desviación de la mirada hacia arriba, lenguaje incoherente, aversión a los símbolos religiosos y demanda de ser llamada por otros nombres. Resultado: Los síntomas se manifestaron durante episodios de hipotensión ortostática, consecuencia de una neuropatía autonómica desarrollada en el contexto de una recaída de un lupus eritematoso sistémico. Se realizó una tomografía computarizada de emisión de fotón único (SPECT) con 99mTc-HMPAO en posición de pie, en presencia de hipotensión postural y los síntomas descritos, la cual mostró una extensa zona de isquemia en el cerebro izquierdo. El mismo estudio se realizó en posición de decúbito, con presión arterial y con un estado mental normal, y fue informado sin alteraciones. Conclusión: Se describe una causa orgánica, de tipo vascular-cerebral, de síntomas similares al de un estado de posesión.

Palabras clave: trastorno del comportamiento, circulación cerebrovascular, lupus eritematoso sistémico, hipotensión ortostática.

Introduction

A constellation of symptoms and signs have been recognized since ancient times and in different cultures as a state of “possession”. There are different points of view to understand the phenomena; for instance, religious, esoteric or medical. The experience of being “possessed” by another entity, such as a person, spirit, god or demon is classified medically as a dissociative (conversion) disorder (International Classification of Disease [ICD-10]) (1). Different theories have been proposed. Special interest has been paid to history of childhood abuse or trauma, suggesting that dissociation may be a psychological defensive mechanism that persists into adulthood. Neuropsychological or purely organic theories have not been demonstrated. In this case report we present a female patient with systemic lupus erythematosus (SLE) with a recurrent possession-like state concurrent with cerebral ischemia induced by episodic orthostatic hypotension associated to autonomic neuropathy. A functional biological explanation for this syndrome is described.

Case Report

A 17-year-old, catholic adolescent female was admitted because cough, progressive dyspnea and chest pain of twenty days of evolution. She denied fever, chills, joints pain, cutaneous, gastrointestinal or genitourinary symptoms. She had a five year history of SLE with symptoms of depression, seizure disorder, type IV nephritis, pancytopenia, polyarthritis; with high titles of antinuclear, anti-Sm and anti-RNP antibodies; an decreased levels of C3 and C4 fractions of complement. At a very early disease stage high-dose of steroid, intravenous cyclophosphamide, rituximab and mycophenolate mofetil were necessary to reach disease control. Then, steroid dose was gradually reduced until its withdrawal a year prior to admission.

On examination, patient had blood pressure 120/70 mmHg,
pulse of 78 beats per minute. The cardiopulmonary system was normal. The abdomen was normal. The joints and skin were with no abnormalities. Neurological evaluation was normal. Laboratory-test results are shown in Table 1. Thorax X-ray and high-resolution CT showed bilateral pulmonary nodules up to one cm on both lungs lobes. A bronchoscopy with bronchoalveolar lavage was made with normal results and infectious diseases were disregarded. Then a lung biopsy by thoracoscopy was done and reported as acute lupus pneumonitis. Serum autoantibodies were reported as follow: positive test for antinuclear antibodies (titer, 1:2560), positive test for anti-dsDNA (titer, 1:320), antcardiolipin-antibody IgM level was highly raised, at 46.8 GPL units, and the antcardiolipin-antibody IgM level was also high, at 29 MPL units. Then, a diagnosis of reactivation of SLE was done, and 1 mg/kg of prednisolone was started. On day 7 of admission, the patient had episodes of aggressiveness. The neurological exploration remained normal. Newly laboratory test were done and infections, metabolic o

<table>
<thead>
<tr>
<th>Variable</th>
<th>Reference</th>
<th>Values</th>
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<tbody>
<tr>
<td>Hematocrit (%)</td>
<td>36.0-46.0 (women)</td>
<td>36.2</td>
</tr>
<tr>
<td>Hemoglobin (g/dl)</td>
<td>12.0-16.0 (women)</td>
<td>12.1</td>
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<td>White-cell count (per mm³)</td>
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<td>Neutrophils (%)</td>
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<tr>
<td>Lymphocytes (%)</td>
<td>22-44</td>
<td>21</td>
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<tr>
<td>Monocytes (%)</td>
<td>4-11</td>
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</tr>
<tr>
<td>Eosinophils (%)</td>
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<td>Basophils (%)</td>
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<tr>
<td>Platelet count (per mm³)</td>
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<tr>
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<td>12</td>
</tr>
<tr>
<td>Partial-thromboplastin time (sec)</td>
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<td>Glucose (mg/dl)</td>
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<td>Sodium (mmol/liter)</td>
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<td>Chloride (mmol/liter)</td>
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<td>Urea nitrogen (mg/dl)</td>
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<td>8.3</td>
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<td>C4 (mg/dl)</td>
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toxic conditions were excluded. Brain Magnetic Resonance Imaging (MRI), was performed revealing hypointense lesions on T2 sequences at frontal and periventricular area. This finding was interpreted as unspecific cerebral involvement related to her prior crisis. Lumbar puncture was performed; the opening pressure was 15 cm of water. The cerebrospinal fluid was xanthochromic and a normal cytchemical analysis was reported. Gram negative, KOH negative, BK negative and cryptococcal and syphilis test were negatives.

Psychiatric assessment was requested. Two concurrent diagnoses were done: a major depressive disorder and a multifactorial delirium. Citalopram 20 mg per day was given and management of underlying organic disarrangement was suggested to control delirium. Patient continued with episodes of psychomotor agitation, aggressiveness, and feeling of dizziness, headache and sialorrhea. The neurological examination was normal. Then quetiapine 50 mg per day was given. On day 10, the psychiatric symptoms got worst, with episodes of disorientation, aversion to religious symbols (which she spat and threw to the ground, requiring their withdrawal of the room), aggressiveness with the family, demanding to be called by another names. She also spoke in inconsistent language during the episodes (listen to the AUDIO). The episodes were more prolonged, which lasted between one and four hours. Between episodes, the patient was asymptomatic with amnesia of misbehavior outbursts. A seizure disorder was suspected but a normal EEG was reported.

It was noticed that symptoms got worst when patient went to bed to the bathroom, so a decision was made to monitor blood pressure and pulse both during lying down and standing positions. Approximately two minutes after standing her arterial pressure drop from normal to 40/20 mm Hg or less and the neuropsychiatric symptoms appear; including movements of right limbs, deviation of the eyes upward, visual disturbances, and incoherent language.

Provocative upright test with single photon emission computerized tomography (SPECT) with 99mTc-D,L-Hexamethylene-propyleneaminoexime (99mTc-HmPAO) was done, detecting abnormal patterns of cerebral perfusion in extensive left brain area. During the application of radiotracer the patient was under hypotension and with ongoing described symptoms. These images were compared with the ones taken during the asymptomatic stage while she was in lying position and normal blood pressure, which showed normal patterns of perfusion in both brains (see Figure 1a y 1b).

All psychiatric medication was withdrawn. On day 14, patient still had orthostatic hypotension, and her pulse did not show variations in frequency. Heart rate response to Valsava maneuver was abnormal.
These were the reasons enough to consider an autonomic disorder, possibly due to SLE reactivation. Pulses of methylprednisolone one gramme daily for three days, a new cycle of rituximab and mycophenolate mofetil three grammes daily were indicated. Fludrocortisone was prescribed with progressive improvement of arterial hypotension and associated symptoms. On day 20, the patient was discharged without orthostatic hypotension, with normal neurological and psychiatric examination.

**Discussion**

The neuropsychiatric systemic lupus erythematosus (NS-SLE) includes several neurologic and psychiatric syndromes when other causes have been excluded. Neurological manifestations include central, peripheral and autonomic syndromes. These manifestations have been reported in up to 60% of patients (2). The cerebrovascular involvement has been found in 5.6% cases (3). Different types of psychiatric syndromes have been described.
and included psychosis, depression, acute confusional state, anxiety, cognitive dysfunction and mood disorder. The patient of this report had NS-SLE syndrome with two sets of symptoms, the initial episode five years before with psychosis, seizures and depression, and the current episode with an autonomic neuropathy. It also describes the development as something novel atypical psychiatric symptoms associated with hypotension and cerebral vascular deregulation secondary to autonomic dysfunction.

The image studies usually requested for NS-SLE are anatomic or functional. With regard to the anatomical studies, the cerebral computerized tomography (CT) and MRI are usually available. The most commonly abnormalities found in CT are generalized or perisurcal atrophy, cerebral infarction and intracerebral haematomas (4,5). Calcifications of diverse structures have been reported rarely (6). With development of MRI, the anatomic information has improved even more; and currently it is considered the standard technique to evaluate morphological brain abnormalities in NP-SLE (7). When it comes to functional techniques to study, for example, the brain perfusion, the positron emission tomography), SPECT and localized proton magnetic resonance spectroscopy are available (8-10).

The study of the brain with Tc99m-HMPAO SPECT in the context of our case was useful in order to demonstrate the acute defects of brain perfusion induced by hypotension in the setting of neurological dysautonomy. Reports from the medical literature in these aspects are poor.

Whether cerebral blood flow autoregulation is maintained in autonomic dysfunction has been debated for a long time. In order to determine how regional cerebral blood flow would change during standing position in patients with idiopathic orthostatic hypotension, Tc-99m HMPAO SPECT studies were performed during postural testing in five patients. After 10 minutes of quiet rest in bed, the patients arose quickly and, at the same time, the radiotracer injected intravenously. Another dose of Tc-99m HMPAO was injected with subjects in the supine position, and SPECT was performed again. In all patients, the authors observed a decrease of blood flow in the frontal cortex and basal ganglia (11).

The correlation between symptoms and signs associated with hypotension and its changes in the brain perfusion was reported in a 78-year-old right-handed man with idiopathic orthostatic hypotension who presented episodes of recurrent stereotyped attacks of bilateral limb shaking and metamorphopsia, which were precipitated by standing for more than 3 or 4 minutes, or walking a few meters. These symptoms would resolve upon squatting or lying down and did not occur.
spontaneously at rest. He did not lose consciousness during the attacks. A single-SPECT showed a moderate compromise of perfusion of the left internal carotid artery territory. After managing orthostatic hypotension his symptoms dramatically disappeared (12).

In this work we present a patient with a recent established autonomic neuropathy evident with postural hypotension which in turn generates neuropsychiatric symptoms. The autonomic neuropathy has been described as a manifestation of NS-SLE and autoimmune pathogenesis had been proposed (13,14).

This paper presents several interesting considerations: First, it shows a form of cerebral ischemia associated to orthostatic hypotension in the setting of autonomic neuropathy in a patient with NS-SLE. Second, it shows a correlation between symptoms and signs of cognitive dysfunction and ischemic findings in representative brain areas with a provocative upright test with 99Tc-99m HMPAO brain SPECT. And finally, it shows how the symptoms and signs of a state of “possession-like” may occur as a result of organic dysfunction, in this case of cerebral vascular type.

References


Cañas C., Echeverri A., Caicedo A., Pabón M., Quiñonez J., Rincón-Hoyos H.

Conflictó de interés: los autores manifiestan que no tienen ningún conflicto de interés en este artículo.

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