Transverse Myelitis in a patient with infertility

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Abstract

Systemic lupus erythematosus (SLE; lupus) is an autoimmune disease. The common neurological manifestations are psychosis, cognitive dysfunction, and headache. Transverse myelitis (TM) may be the first manifestation of lupus. We describe one such case in which TM was the only presentation of SLE.

Covering letter

Dear Editor-in-Chief

Of the Clinical Case Reports

We would like our manuscript entitled "Transverse Myelitis in a patient with infertility" to be considered for publication in the Clinical Case Reports. All authors have contributed significantly and are in agreement with the content of the manuscript. This is an original research article that has not been published elsewhere, and all authors declare no conflict of interest related to the manuscript. This manuscript has been prepared according to the journal instructions.

Thank you for considering this manuscript for publication.

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Key clinical message:

More attention from the medical staff to this sign can lead to faster diagnosis and timely treatment of patients.

Keywords: Systemic lupus erythematosus, SLE, Transverse Myelitis, Longitudinally extensive spinal cord lesion

Abbreviations:

SLE: Systemic lupus erythematosus

TM: Transverse myelitis

M/F: Muscle force

MRI: Magnetic resonance imaging

VEP: Visual Evoked Potential

LESCL: Longitudinally extensive spinal cord lesion

Glu: Glucose

Pro: Protein

LDH: Lactate dehydrogenase

WBC: White Blood Cells

RBC: Red Blood Cells

LP: Lumbar puncture

DVT: Deep vein thrombosis

ANA: Anti-nuclear antibody

dsDNA: Anti-Double-Stranded deoxyribonucleic acid

CTA: Computed Tomography Angiography

PTE: Pulmonary Thrombo-Embolism

ATM: Acute Transverse Myelitis

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Conflict of interest:

None

Ethics

This study was performed according to the principles outlined by the World Medical Association's Declaration of Helsinki on experimentation involving human subjects, as revised in 2000, and has been approved by the ethics committee of the Tabriz University of Medical Sciences.

Author Contributions:

Amirreza Khalaji: corresponding author and conception and design of the report and final approval of the version to be submitted.

Alireza Khabbazi: Final approval of the version to be submitted.

Leyla Ghadakchi: Revising the article critically for important intellectual content.

Yalda Yazdani: Drafting the article or revising it critically for important intellectual content.

Elham Habibzade: Drafting the article or revising it critically for important intellectual content.

Saba Mehrtabar: Data collection.

Consent statement:

Written informed consent was obtained from the patient to publish this report and clinical images. Consent has been signed and collected in accordance with the journal's patient consent policy.

Introduction

Systemic lupus erythematosus (SLE; lupus) is an autoimmune disease that impacts public health, and in females between five to sixty-four years is between the top 20 main causes of mortality (1). SLE manifestations correlated with subsequent immune complex deposition and formation, various autoantibodies, and other immune processes (2, 3). The neurological manifestations are seen in the 25-95% of cases with SLE. Common manifestations include psychosis, cognitive dysfunction, and headache (4, 5). According to one research, most patients have active lupus symptoms that precede transverse myelitis (TM); however, TM may be the first manifestation of lupus (6). TM is a spinal cord inflammatory condition that may present acutely or sub-acutely (7). It is an uncommon condition with an incidence of approximately one to four new cases per million individuals every year (8). TM is histologically defined by immune cell infiltration of the spinal cord, with pathogenesis mediated by various immunological pathways (9). We describe one such case in which TM was the only presentation of SLE.

Case Presentation

The patient is a 40-year-old non-smoker female with a 17-year history of infertility. She previously underwent hormone therapy; she developed acute lower extremity weakness around six years ago, predominantly on the right side. Her family history was negative for any kind of connective tissue disorders or SLE. Additionally, the patient had a history of severe headaches and flutter, for which she was referred to a neurologist. She was admitted to the neurology ward because of her acute presentation. During hospitalization, the initial examination determined a decrease in Muscle Force (M/F) with a predominance for the right side, a positive Marcus Gan test in the left eye, and the presence of a sensory level in the C8 and T1 dermatomes, and abnormal cerebellar tests.

Meanwhile, his other vision examinations were intact. According to the clinical findings, the visual evoked potential (VEP) requested for her was normal, and Magnetic resonance imaging (MRI) showed multiple plaques of the brain and a longitudinally extensive spinal cord lesion (LESCL). Glucose (Glu): 49mg/dl, Protein (Pro): 28mg/dl, Lactate dehydrogenase (LDH): 34IU/L, White Blood Cells (WBC): 0 cells/μL, and Red Blood Cells (RBC): 0 cells/μL, and there was no evidence of Oligoclonal banding reported in the Lumbar puncture (LP) done for her suspicious demyelinating disorders. Also, she developed Deep vein thrombosis (DVT) in her right upper extremity.

Following a partial recovery, the patient was discharged with a diagnosis of transverse myelitis in demyelinating disease. Due to her relatively complete recovery over the last two years, the patient has arbitrarily discontinued her medication and follow-up. The patient presented with acute weakness in the lower extremities about a month ago and was hospitalized in the neurology ward. Her blood pressure was 115/78, her pulse rate was 83 beats per minute, and her temperature was 36.6°C. Lung, heart, and abdomen examinations were unremarkable. M/f was 4.5 for the right upper extremities, 5.5 for the left upper extremities, and 0.5 for the lower extremities during the initial examination; Marcus Gunn was negative, and tendon reflexes were +3 for the right knee and +2 for the left knee. TM was detected during this hospitalization via MRI. The patient had 12 doses of corticosteroids over six days and then six plasmapheresis sessions, which did not considerably improve her general health. Complement depletion, platelet depletion, proteinuria, positive Anti-nuclear antibody (ANA), and anti-Double-Stranded deoxyribonucleic acid (dsDNA) antibodies were detected in lab testing for suspicions Rheumatic diseases. During hospitalization, the patient had shortness of breath and was assessed for pulmonary artery Computed Tomography Angiography (CTA) and echocardiography. Pulmonary Thrombo-Embolism (PTE) was proposed in the CTA of her pulmonary arteries. In the echocardiogram done for her, a lesion on the heart valves was seen, suggesting Lyman Sachs endocarditis, and pericarditis was reported. They suspected SLE and APS disorders according to the patient's clinical condition and imaging and testing results. The patient was referred to the rheumatology ward. Her clinical diagnosis of lupus was validated through clinical trials and diagnostics—the rheumatologist prescribed Rituximab one gram every two weeks, which dramatically improved the patient's clinical condition. The patient is in good general condition and has stable vital signs in subsequent follow-ups.

Discussion

The most prevalent SLE neurological symptoms are seizures, psychosis, or headache. 1-2 percent of SLE patients' Acute Transverse Myelitis (ATM) is seen, while in one adult study, up to 39 percent of subjects are presenting features (4, 10, 11). S. Zhang et al. (12) demonstrated that patients over the age of 40 years were more likely to have developed severe myelitis initially, explaining why younger individuals have a poor prognosis. Moreover, extensive spinal cord lesions, initial severe neurological impairment, delayed steroid impulse therapy, and hyper-inflammation could predict a poor prognosis of systemic lupus erythematosus with Transverse Myelitis. Meanwhile, another study shows that Transverse myelitis may present as a symptom of SLE or develop more than ten years after diagnosis of SLE. The primary prognostic factor is the severity of the initial neurological flare (with paraplegia). Thus, ATM signs and unexplained sensory complaints should elicit additional examinations in patients with SLE (13).

The majority of cases who develop ATM complications do so within five years after being diagnosed with SLE. The classical presentation of ATM may occur in SLE patients, such as sphincter disturbances, motor weakness, and sensory disturbance. Our patient did not show typical SLE or transverse myelitis symptoms.

Our patient was diagnosed with SLE according to the presence of positive immunological markers and an atypical clinical presentation of longitudinal myelitis. Although the mechanism by which ATM causes SLE is unknown, it is most likely caused by immune complex-mediated thrombosis or vasculitis, which results in ischemic spinal lesions, or by anti-phospholipid antibodies cross-reacting with spinal cord phospholipids (11, 14). In SLE patients where ATM is presenting manifestation, numerous patients probably do not fulfill SLE diagnostic criteria; however, during the disease, they may develop further signs and symptoms of SLE in the future (4). Although the ANA and double-stranded DNA antibodies were positive in our reported case, the patient showed no additional clinical symptoms associated with SLE. The patient, after aggressive therapy, showed reasonable improvement.

Steroids in Intravenous pulse methylprednisolone and immunosuppressant such as cyclophosphamide are used in the general therapy strategy (10, 15). This combination of therapies appears to have a more favorable outcome. However, the prognosis is often seen as bad in patients with SLE. Plasmapheresis has been utilized in some patients, although its role is unclear (14). Although most SLE-associated ATM patients have satisfactory neurologic outcomes following high-dose corticosteroid therapy, some patients have a recurrence and permanent neurologic deficits (16).

In conclusion, this study shows that ATMs may, on rare occasions, represent the initial manifestation of SLE. ATM secondary to SLE, despite improved treatment methods, can have a bad prognosis and requires more than only steroids. Early detection and aggressive treatment can help prevent long-term irreparable damage and may positively result. This article underlines the importance of multicenter studies and establishing a registry for individuals with SLE who have ATMs to aid in the study of best management choices.

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