Clinical Medicine Insights: Case Reports

CASE REPORT

Chorea as a First Manifestation in Young Patients with Systemic Lupus Erythematosus Who Was Initially Diagnosed With Rheumatic Fever

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Abstract: Chorea is a rare manifestation of systemic lupus erythematosus (SLE). We report on a young patient with chorea who was diagnosed initially with rheumatic fever. Follow up and further evaluation confirmed the diagnosis of SLE and anti-phospholipid syndrome. Of special interest were the negative antiphospholipid (aPL) antibodies and the initial diagnosis of rheumatic fever which is still not uncommon problem in our region. The rarity of such presentation with joint and non specific increase of antistreptolysin O (ASO) titer might be the factors that led to an incorrect diagnosis. Early diagnosis and treatment of SLE and anti-phospholipid syndrome are very crucial and should be considered with such presentation.

Keywords: lupus anticoagulant, anticardiolipin antibody, thrombosis, systemic lupus erythematosus, connective tissue disease

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Introduction

Systemic lupus erythematosus (SLE) affects many organ systems, including the central and peripheral nervous systems and muscles.[1,2] Movement disorders were not included in the American College of Rheumatology (ACR) criteria.[3] Chorea has been seen in fewer 4% of patients.[4] In this case report, the chorea was the first manifestation in this SLE patient.

Case Report

A 15-year old right-handed female was presented with a two day history of involuntary movements affected the right side of her body, which had been noted primarily by her mother. She also had a history of multiple joint pain and of upper respiratory tract infection two weeks prior to her symptoms where she received 7 days of Amoxicillin. She had denied any history of fever or shortness of breath. She was not on any medication. There was no family history of movement disorders, dementia or psychiatric illness.

The patient reported no difficulty with speech, balance or autonomic function, and there was no history of psychiatric illness or cognitive deterioration. There was also no history of involuntary movements during childhood or pregnancy, and the patient was not aware of any past history of rheumatic fever.

General physical examination was normal apart from mild fever (37.7 °C) and the presence of joint tenderness of the knees, hands and feet. Neurological examination revealed normal extra-ocular movements, and normal motor, sensory and cerebellar testing. Reflexes were physiological and symmetrical. The patient's choreic movements were moderate in severity, and predominantly distal, affecting the right arm and leg. The cardiovascular system and skin examination were unremarkable.

Investigations including renal and liver function, complete blood count were all unremarkable. Antistreptolysin O (ASO) titer was mildly elevated, ECG & echocardiogram was normal. Brain MRI was normal. Erythrocyte sedimentation rate, activated partial thromboplastin time, as well as levels of glucose, thyroid-stimulating hormone, thyroid antibodies, electrolytes, calcium, magnesium, phosphorus were normal.

The patient was admitted to the hospital with working diagnosis of rheumatic fever. She was treated with salicylates, single dose of intramuscular benzathine penicillin G and Haloperidol.

Two weeks later, throat swab culture, serum vitamin E and ceruloplasmin, were all unremarkable but involuntary movements persisted. Therefore, Antinuclear Antibody (ANA) and anti-double-stranded (ds) DNA antibody titers were carried out which were found to be elevated (1/640; 36.5 IU/mL, normal $<$ 10 IU/mL respectively). The anti-Sm and anti-cardiolipin (aCL) antibodies were both negative. Repeated ASO titer remained the same as before.

At this stage, because the results of investigations were highly suggestive of SLE, 24 hours urine collection of proteins showed 2300 mg/day, kidney biopsy was done and showed glomerulonephritis (WHO class 4). At that time, she was started on Cellcept 2 gm/day and prednisolone.

Six months after treatment, she had complete remission of lupus nephritis but remained with mild choreo-athetotic movement. The repeated IgM aCL antibodies were positive.

Discussion

Rheumatic fever is the most common cause of acquired chorea in the young.[5,6] No single specific laboratory test can confirm this diagnosis.[7–9] The modified Jones criteria which are less sensitive tend to make cases more easily missed in high-prevalence areas.[10,11] This is why this patient was treated as having rheumatic fever. High titer of ANA on this patient alerted the doctors that SLE should be considered as a cause of such presentation.

Chorea, although rare, is a well recognized neurological manifestation of SLE.[12,13] It usually occurs during the course of disease but may be the presenting feature of SLE.[14] In the retrospective study of central nervous system disease in 105 patients with SLE, Khamashta et al[15] observed two patients with chorea as a presenting feature of their illness. In this case report, chorea was also the initial manifestation of SLE, though misdiagnosed as rheumatic fever.

Most reports show an association between chorea and antiphospholipid (aPL) antibodies in SLE patients.[16,17] In this patient, the aPL antibodies were negative initially but turned out to be positive six months after presentation. The aPL antibodies are unlikely to be positive in patients with rheumatic

fever as previously mentioned by Asherson and associates.[18]

The chorea in SLE is immune-mediated problem and associated with appearance of lupus anticoagulants and aCL antibodies in the serum.[19] Even normal titers of these antibodies at the time of presentation, however, do not rule out the disease. Baizabal-Carvallo and colleagues[16] in their retrospective review initially observed negative aPL at the onset of chorea. Later on, IgM aCL antibodies became positive. This case report also reveals elevated aCL antibodies six months after the occurrence of chorea in SLE.

Conclusion

The non-sensitive criteria of Rheumatic fever should make the physician alert to other possible causes of chorea like SLE and antiphospholipid syndrome. The aPL antibodies should be repeated periodically even if it is negative at the time of presentation.

Disclosures and Ethics

As a requirement of publication author(s) have provided to the publisher signed confirmation of compliance with legal and ethical obligations including but not limited to the following: authorship and contributorship, conflicts of interest, privacy and confidentiality and (where applicable) protection of human and animal research subjects. The authors have read and confirmed their agreement with the ICMJE authorship and conflict of interest criteria. The authors have also confirmed that this article is unique and not under consideration or published in any other publication, and that they have permission from rights holders to reproduce any copyrighted material. Any disclosures are made in this section. The external blind peer reviewers report no conflicts of interest.

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