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CHOREA AS A RARE MANIFESTATION OF NEUROPSYCHIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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ABSTRACT
Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with multisystemic involvement. It has variable clinical presentations and a relapsing and remitting course. Clinical manifestations are broad, ranging from mild mucocutaneous involvement to major organ involvement. We present a case of a 32-year-old female with repeated movements of the right arm, and confusion with a background of pains in multiple joints. She was diagnosed as a case of Neuropsychiatric Systemic Lupus Erythematosus (NPSLE). She was managed with pulse therapy of cyclophosphamide (CYC) along with other supportive medications. After six months of intensive immunosuppressive therapy, she remained well and continued maintenance therapy.

KEYWORDS: Chorea, Systemic lupus erythematosus, ANA, Anti-Smith

INTRODUCTION
Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder primarily affecting women, typically starting at childbearing age. It has variable clinical manifestations including rash, arthritis, and fatigue.[1] Severe cases may present with nephritis, neurological complications, anemia, and thrombocytopenia. Most patients have positive anti-nuclear antibodies (ANA). Neuropsychiatric lupus (NPSLE) is seen in about 20% of cases.[2] NPSLE has different clinical features depending on central and peripheral nervous system involvement. Our patient had confusion and chorea as rare features of NPSLE, along with other musculoskeletal features and immunological evidence of SLE.

CASE PRESENTATION
A 32-year-old female resident of Jhelum presented to the OPD of District Head Quarters Hospital, Jhelum, with a history of excessive movements of her right arm for two days. These movements were unintentional and irregular. She was not able to perform hand grip or any voluntary act with her right arm due to these movements. She also gave a history of clumsiness and getting confused for the last one month. She denied headache, vomiting, or high-grade fever. On further inquiry, she reported experiencing pain and mild swelling in the small joints of her hands, shoulders, and knees for the past five months. The joint pain was symmetrical, of mild to moderate intensity, and accompanied by morning stiffness lasting more than 30 minutes.. She mentioned having a low-grade fever, fatigue, and rash over the face. She had suffered from significant hair loss. She did not report dyspnea, pedal edema, cough, or frothy urine, and there was no history of bluish discoloration of the fingertips suggestive of Raynaud's phenomenon.

On examination, she was a young lady, well-oriented, but she was slow to respond and confused. She had a pulse rate of 80 bpm, blood pressure 110/70 mmHg, temperature 97.8 °F, and respiratory rate 18 breaths/min. She had excessive movements of the right arm, these movements involved both the proximal and distal arm, but the distal muscle groups were more affected. Movements were spontaneous, jerky, irregularly timed and repetitive. These were randomly distributed, and flowing in character. Motor impersistence was observed; she could not maintain a steady hand grip (milk maid grip). However, other parts of the body did not show motor impersistence. These movements were observed only in the right arm and the rest of the body was spared.

She had a rash over her cheeks, sparing nasolabial folds. She had oral ulcers and sparse skull hairs. She had no deformities of the small joints of her hands. There was no motor weakness, cerebellar dysfunction or any cranial nerve involvement. The rest of the systemic examination was unremarkable.

She was admitted to the medical ward. Her laboratory test reports are shown in Table 1. CT scan of Brain

(plain) was normal. Diagnosis of NPSLE was made. Chorea was treated with haloperidol 5mg TDS, which resulted in significant improvement. Specific treatment started with IV cyclophosphamide (CYC) along with methylprednisolone. Choreiform movements subsided after the first dose of CYC. Pulse therapy was given for six months with monthly CYC and methylprednisolone. Home medication included azathioprine, proton pump inhibitors, prednisolone 7.5mg OD, vitamin D3, and calcium supplements. After six months of immunosuppressive therapy, she had significant improvement in other symptoms like hair loss, photosensitivity, and mouth ulceration.

Table 1: Lab test reports

Hb	12 g/dl
WBC	9000/μL
PLT	203,600/μL
LFT	Br 1 mg/dl, ALT 35 IU/L, AST 38 IU/L, Alk. Phosph112 IU, Albumin 4.2 gm/dl
RFT	Creatinine 0.7 mg/dl Urea 40 mg/dl
ELECTROLYTES	Na 134 meq/L K 4,1 meq/L
Serum calcium	9.2 mg/dl
Serum phosphate	3.8 mg/dl
HEP B,C	Negative
HIV	Negative
Urine R/E	Absent proteins, RBCs or Cast
Urinary protein/creatinine	0.20 (normal value Up to 0.20)
RA Factor	59.6 (> 14 positive)
TSH	2.2 mU/L(0.4 - 4)
CRP	3mg/dl(0-5)
ESR	20 mm/1 hour
Anti CCP	Result: Negative < 8 (neg <17, pos>17),
ANA	Positive
Anti dsDNA	Result: Negative Value: 0.1 (neg <5, pos >10),
Anti Smith	Positive
C3 C4	26(90-180 mg/dl) 7(15 -57 mg/dl)
Anti-cardiolipin IgM	1.0 (pos >7)
Anti-phospholipid IgM	1.6 (pos >10)
Beta 2 glycoprotein IgM	2.1 (pos>8)

DISCUSSION

SLE, an autoimmune condition characterized by relapses and remissions, affects individuals worldwide, particularly women of childbearing age. Women aged 15 to 44 have a female-to-male ratio of up to 13:1.[3] SLE is characterized by the production of autoantibodies and the formation of immune complexes, triggering inflammatory responses that lead to organ damage. In mild SLE, patients may have only constitutional, mucocutaneous, or musculoskeletal involvement. While in moderate or severe SLE, there is involvement of other organs such as the kidneys (renal lupus), the nervous system (neuropsychiatric lupus), serosal surfaces and the hematologic system. The EULAR classification helps clinicians in making diagnosis of SLE, ensuring appropriate management of its diverse clinical presentations.[3]

NPSLE consists of various neuropsychiatric symptoms directly linked to SLE. The American College of Rheumatology (ACR) outlines 19 specific clinical syndromes of NPSLE, such as cognitive impairment, seizures, demyelinating syndrome, headaches, strokes, and movement disorders.

Chorea is defined as "a state of excessive, spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character. These movements can range in severity from mild restlessness with exaggerated gestures and expressions, fidgeting of the hands, and a dance-like gait, to severe, continuous, violent movements. The term "chorea" is derived from the resemblance of these movements to dancing. The underlying pathology generally involves dysfunction within the basal ganglia. While chorea is classically associated with Huntington's disease, it can also occur in variety of other conditions. Chorea is classified as "primary" when linked to Huntington's disease (HD) or other genetic causes, and "secondary" when associated with factors such as infections, medications, metabolic issues, autoimmune disorders, or paraneoplastic syndromes.[4] The key approach to managing secondary chorea is addressing the underlying condition causing it. Chorea is the most common movement disorder observed in SLE.[5] It typically appears early in the course of SLE and can even be the first symptom. Chorea is strongly linked to antiphospholipid antibodies (aPL), which are present in 25% to 35% of SLE patients.[6] While there are no specific MRI findings for SLE-related chorea, some cases may show ischemic damage in the basal ganglia.

Treatment for NPSLE typically entails immunosuppressive therapy combined with corticosteroids. Azathioprine and cyclophosphamide are particularly effective immunosuppressants in managing NPSL. The standard treatment for severe or life threatening SLE, consist of high-dose glucocorticoids and intravenous (IV) pulse CYC, based on a series of randomized controlled trials (RCTs) conducted by the National Institutes of Health (NIH) .There are different CYC regimen , NIH recommends monthly high dose CYC (0.5 to 1g/m2) for six months along with high dose methylprednisolone.[7]

CONCLUSION

Chorea is a rare manifestation of neuropsychiatric systemic lupus erythematosus that requires timely recognition and prompt immunosuppressive therapy for optimal outcomes. This case highlights the importance of considering NPSLE in atypical neurological presentations to ensure early diagnosis and effective management.

REFERENCES

1. Justiz Vaillant AA, Goyal A, Bansal P, Varacallo M. Systemic Lupus Erythematosus [Internet]. PubMed. Treasure Island (FL): StatPearls Publishing; 2021. Accessed on July 7 2025. Available from: https://www.ncbi.nlm.nih.gov/books/NBK535405
2. Ameer MA, Chaudhry H, Mushtaq J, Khan OS, Babar M, Hashim T, et al. An Overview of Systemic Lupus Erythematosus (SLE) Pathogenesis, Classification, and Management. Cureus. 2022 Oct 15;14(10).
3. Fava A, Petri M. Systemic lupus erythematosus: Diagnosis and clinical management. J Autoimmunity. 2019 Jan;96(1):1–13.
4. Feinstein E, Walker R. Treatment of Secondary Chorea: A Review of the Current Literature. Tremor Other Hyperk Movements. 2020;10(1).
5. Sharma R. Systemic lupus erythematosus (CNS manifestations) | Radiology Reference Article | Radiopaedia.org [Internet]. Radiopaedia. Accessed on July 1 2025. Available from: https://radiopaedia.org/articles/systemic-lupus-erythematosus-cns-manifestations?lang=us
6. Aringer M, Costenbader K, Daikh D, Brinks R, Mosca M, Ramsey-Goldman R, et al. 2019 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus. Arthritis Rheumatol. 2019;71(9):1400-12.
7. Mok CC. Con: Cyclophosphamide for the treatment of lupus nephritis. Nephrology Dialysis Transplantation. 2016 May 14;31(7):1053–7.

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