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Unmasking Lupus: Chorea as the Sole Initial Manifestation

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Abstract **Case Report**

Systemic lupus erythematosus (SLE) is an autoimmune disease with a wide spectrum of clinical manifestations, including neuropsychiatric systemic lupus erythematosus (NPSLE). Although a recognized feature of NPSLE, Chorea is an uncommon initial presentation, particularly in children. We report the case of a 12-year-old girl who presented with acute generalized chorea as the first manifestation of SLE. Extensive evaluation, including clinical, laboratory, and imaging studies, confirmed the diagnosis of SLE and identified neuropsychiatric involvement. This case underscores the importance of considering SLE in the differential diagnosis of pediatric chorea and highlights the diagnostic and therapeutic challenges in managing NPSLE with chorea as the primary feature.

Keywords: SLE, Autoimmune disease, Chorea, NPSLE, Adolescent.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disorder, characterized by the presence of autoantibodies, systemic inflammation, and multiorgan involvement, including, less frequently, the central and peripheral nervous system [1]. The clinical presentation of the condition varies widely. The classification of SLE is established based on specific criteria, which include only psychosis and seizures as nervous system manifestations; nevertheless, also other neuropsychiatric disorders are associated with SLE (neuropsychiatric SLE) [2]. Neuropsychiatric symptoms of SLE (NPSLE) are particularly difficult to identify and diagnose but account for a considerable percentage of manifestations, with an estimated prevalence ranging from 20% to 95% of patients with pediatric SLE [1,3].

The American College of Rheumatology released a proposal for the definition and nomenclature of NPSLE in 1999 [4]. Movement disorders were one of the 19 syndromes described in the proposal. Chorea is a hyperkinetic movement disorder characterized by non-repetitive, abrupt, involuntary jerky movements that may be unilateral or generalized. The movements are non-patterned with variable speed, timing, and direction, flowing from one body part to another and giving, in less severe cases, an appearance of fidgetiness. The unpredictable nature of chorea distinguishes it from tremor and dystonia [5]. Chorea has numerous etiologies, including structural, pharmacologic, autoimmune, metabolic, and genetic. Chorea, although rare, is well described in SLE, with a cumulative incidence of 0.6% [6].

Even if the most common etiology of chorea in pediatric patients is the autoimmune form of post-streptococcal origin, it is a well-recognized albeit rare neuropsychiatric manifestation of systemic lupus erythematosus (SLE) [7].

In this study, we report a case of a 12-year-old girl who presented with acute generalized chorea as the first manifestation of SLE.

CASE PRESENTATION

A 12-year-old girl in class 6, presented with complaints of generalized weakness and involuntary hand movements persisting for four months. The abnormal movements began insidiously, progressively involving both hands and over the last three months, they extended to the entire body. The involuntary movements are now static in progression. These symptoms were preceded by a history of sore throat and fever, which resolved spontaneously 15 days before symptom onset.

There was no history of joint pain, shortness of breath, body swelling, oral ulcers, photosensitivity, or alopecia.

The patient has a past medical history of tubercular lymphadenitis diagnosed 1.5 years ago, for which she completed Category 1 antitubercular therapy and was declared cured. She is fully immunized as per the childhood immunization schedule but has not received adult vaccines.

She is the fifth child of non-consanguineous parents. Her father, a businessman, earns approximately 20,000 taka per month. The family history is unremarkable for similar illnesses or hereditary conditions.

Initial Examination and Investigations: On general examination, the patient appeared pale, with a pulse rate of 90 beats per minute, blood pressure of 90/60 mmHg, and a body temperature of 98.4°F. Involuntary, irregular movements of both hands were noted, along with a positive *milkmaid grip*. The BCG vaccination scar was present. There was no edema or lymphadenopathy.

- **Cardiovascular Examination**: Normal heart sounds (S1, S2) with no murmurs.
- **Neurological Examination**: Higher cerebral functions, including speech, were intact. Cranial nerve function, motor and sensory systems, and cerebellar signs were normal.

A comprehensive series of laboratory and imaging studies were performed to evaluate the etiology of the symptoms:

Laboratory Investigations: A complete blood count (CBC) test was done on the patient. The findings showed that the patient had a hemoglobin level of 8.19 g/dL, which led to the diagnosis of microcytic hypochromic anemia. ESR was 100 mm at the end of the 1st hour. Total leukocyte count was $6.6 \times 10^3/mm^3$ and Platelet count was $663 \times 10^3/mm^3$. Peripheral Blood Film (PBF) was consistent with microcytic hypochromic anemia. The patient had a serum iron level of 93 μg/dL, total iron binding capacity (TIBC) was 286 μg/dL, and ferritin level was 90.7 ng/mL. All these iron studies were within the normal ranges. Hemoglobin electrophoresis identified the Beta-thalassemia trait in the study patient. Both serum electrolytes and urine routine examination reports were normal. Serum creatinine was 0.6 mg/dL and alanine transaminase (ALT) was 26 U/L. All these findings were normal. Serum ceruloplasmin was 22 mg/dL which was also normal. Antistreptolysin O titer was elevated at 800 IU/mL. C-reactive protein (CRP) test shows that the patient had a normal (1.2 mg/L) level of CRP, which indicates less inflammation in the body.

Autoimmune Profile: Antinuclear antibody (ANA) was positive with ≥2.75 times normal and a fine speckled pattern. Anti-dsDNA was positive as the patient had 110 IU (Positive ≥75 IU). Antiphospholipid antibodies test showed 21 IU which means positive (Positive ≥ 12 IU). Lupus anticoagulant test showed 52 which was positive (Positive ≥36). Activated partial thromboplastin time (APTT) test showed 62.5 seconds which was prolonged (Prolonged time ≥30 seconds). Prothrombin time was mildly prolonged with a test result of 18.1 seconds (Prolonged time ≥15 seconds).

Imaging and Neurological Studies: The magnetic resonance imaging (MRI) was normal and the electroencephalogram test says that the patient had normal electrical activity of the brain

Cardiac Evaluation: Electrocardiogram (ECG) was found normal in our patient. The findings of the 2D Echocardiogram were also normal.

Management & Follow-up: The patient showed significant improvement within a month of starting hydroxychloroquine (5mg/kg), Ecosprin (aspirin), haloperidol (low dose), and Perkinil (trihexyphenidyl). The inclusion of haloperidol and Perkinil suggests the presence of neuropsychiatric symptoms, possibly neuropsychiatric lupus (NPSLE). Haloperidol was gradually withdrawn after four months, reflecting the resolution of acute neuropsychiatric symptoms. Ecosprin and hydroxychloroquine were continued as maintenance therapy. Over the last two years, the patient has remained symptom-free with no flares or recurrence of SLE symptoms, indicating excellent disease control.

Prognosis: The patient's stable remission and absence of SLE flares for two years is a highly encouraging outcome. With continued adherence to treatment and regular follow-up, the long-term outlook remains favorable.

Figure 1: A 12-year-old SLE patient

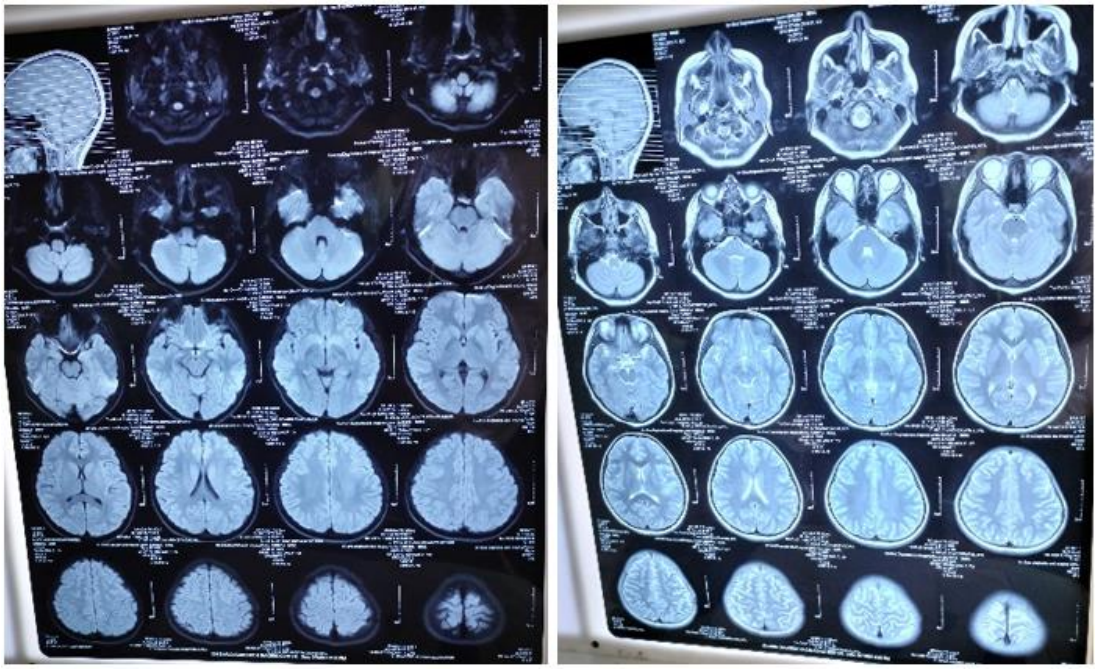

Figure 2: MRI findings of the patient

DISCUSSION

The presentation of generalized weakness and involuntary movements in a child raises a broad differential diagnosis, encompassing neurological, infectious, autoimmune, and metabolic etiologies. In this case, the presence of progressive choreiform movements alongside serological and immunological findings led to the diagnosis of systemic lupus erythematosus (SLE) with neuropsychiatric involvement, specifically chorea.

Neuropsychiatric systemic lupus erythematosus (NPSLE) is a complex and heterogeneous manifestation of SLE, involving the central or peripheral nervous system. It occurs in 25–75% of SLE patients, with symptoms ranging from mild cognitive dysfunction to severe neuropsychiatric syndromes such as psychosis, seizures, and movement disorders like chorea [8,9]. Chorea in SLE is rare, reported in less than 2% of NPSLE cases, and is more commonly observed in children and adolescents with SLE often has an immune-mediated

basis [10]. Autoantibodies, such as antiphospholipid antibodies (APLA), play a key role by disrupting neuronal signaling pathways and causing microthrombi or vascular inflammation in the brain [11]. The presence of APLA in this patient, along with positive lupus anticoagulant and prolonged activated partial thromboplastin time (APTT), supports this mechanism.

The history of sore throat and elevated anti-streptolysin O (ASO) titers also raises the possibility of Sydenham's chorea, a manifestation of acute rheumatic fever [12]. However, this diagnosis is less likely given the absence of major Jones criteria, such as carditis, arthritis, or erythema marginatum. Additionally, the prolonged static phase of chorea, the absence of evidence for recent group A streptococcal infection on further testing, and the presence of hallmark SLE antibodies (anti-dsDNA and ANA) collectively favor NPSLE as the underlying etiology [13].

Antiphospholipid syndrome (APS) is a known comorbidity in SLE patients and is characterized by vascular thrombosis and pregnancy-related morbidity [14]. In pediatric patients, APS can manifest with non-thrombotic neurological symptoms, including chorea. This patient's elevated APLA levels and clinical presentation highlight the overlap between APS and NPSLE. Timely recognition is critical to prevent thrombotic complications and guide anticoagulation therapy if indicated.

In terms of imaging testing, MRI is the most often used examination for these patients because, while it is not specific for the diagnosis of SLE, it allows differential diagnosis for other causes. Global records show hemichorea presentations in stroke patients in white substance, even though chorea is frequently associated with subthalamic nucleus lesions [15], provided that they affect the nucleo-linking pathways. In 40% of NPSLE patients, the MRI is normal and there are no correlations between imaging findings and clinical symptoms or SLE-specific changes [16].

Clinical Implications and Management

This case highlights the significance of considering SLE in children with atypical neurological symptoms, particularly in the absence of common systemic manifestations such as renal involvement or arthritis. Early diagnosis enables prompt initiation of therapy, which may involve immunosuppressive drugs for SLE and anticoagulation in cases of significant antiphospholipid antibody positivity. Multidisciplinary treatment combining pediatric rheumatologists, neurologists, and hematologists is essential to optimize outcomes in such complex presentations.

CONCLUSION

This patient's case illustrates the diagnostic challenge posed by a rare initial manifestation of SLE. The overlap between post-streptococcal syndromes and autoimmune diseases further complicates the clinical picture. Thorough evaluation, including detailed immunological profiling and exclusion of alternative diagnoses, remains crucial in pediatric patients with movement disorders.

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