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Lupus Disguised as Chorea: Uncommon Presentation of a Common Disease

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Bhumika G.C.-writing of draft,editing Shrijana Shrestha-supervision,review Firoz Anjum-investigation Shreya Thapa- data curation Rahul Shrestha-editing

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The authors declare no conflict of interest.

CONSENT

Written informed consent was taken from the patients father in accordance with the journal's policy.

Key clinical message

This case report presents a preadolescent female with a rare presentation of lupus as isolated chorea. An investigation to find out the cause of unexplained cytopenia with raised inflammatory markers ultimately led to the diagnosis of neuropsychiatric lupus with antiphospholipid antibodies. Lupus should always be included in the differential for chorea in children.

Keywords: lupus, chorea, paediatrics, neuropsychiatric lupus.

1 Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease of unknown cause that can affect any organ system. Although SLE in children is fundamentally the same disease as in adults, with similar etiology, pathogenesis, and laboratory findings, there are some differences in frequency and severity of certain clinical manifestations.[1]

In 1999, the American College of Rheumatology issued a proposal for the nomenclature and definition of Neuropsychiatric SLE(NPSLE). The proposal defined 19 syndromes, including movement disorders.[1]Although NPSLE is documented in a range of 13.5% to 34.6% of paediatric patients diagnosed with SLE,[2,3] chorea is a rare neurologic syndrome associated with SLE, described in 1 to 3 percent of patients with SLE. While the predominant cause of chorea in children is typically the autoimmune variant resulting from post-streptococcal origins, it's important to note that chorea can also arise as a complication of Systemic Lupus Erythematosus (SLE).[4]

We report a case of acute onset chorea as a form of NPSLE.

2 Case history

A 12-year-old female presented to the emergency department of a tertiary care centre in Nepal with random dance-like involuntary movements affecting the limbs which gradually increased in frequency and severity over the course of 2 weeks. The movements initially involved her left upper and lower limbs, later affected all four limbs, but were not associated with any changes in the level of consciousness and were notably absent when asleep. Her history was insignificant except for sore throat and fever for a few days, 4 months prior, which resolved without treatment. She did not have any history of altered sensorium, seizures, degrading school performance, loss of consciousness, incontinence, fever, vomiting, headache, joint pain, weight loss, rashes, ulcers, toxin exposure.

On admission, she was a well oriented, well-built prepubertal girl with generalized choreiform movements, a darting tongue, milkmaid's grip, pronator drift and spooning sign. The remaining general and systemic examinations were normal.

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3 Methods

Initial workup revealed bicytopenia (low hemoglobin for her age and leukocytopenia) with a borderline platelet count and an increased erythrocyte sedimentation rate\sout.

Table 1: Results of initial laboratory investigations

Parameters	Values
Total leukocyte count	3590/cubic millimetre
Differential count	70% Neutrophils 30% Lymphocytes (Absolute lymphocyte count 1077)
Haemoglobin / haematocrit	11g/dl/33%
Platelet count	166000/cubic millimetre
Erythrocyte sedimentation rate	87mm/hour
Antistreptolysin O	Negative
Urea/Creatinine	44/0.6 mg/dl
Sodium/Potassium	138/4.3 mmol/litre

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After the initial evaluation, she was started on Haloperidol 0.25mg thrice daily to manage the disabling movement disorder.

The bicytopenia revealed in the laboratory test results led to further workup in the line of an autoimmune disorder which showed an elevated anti-nuclear antibody (ANA) titre of 1:640 (Normal Range(NR) histone antibodies with hypocomplementemia- low C3 and C4 ,C3 39 mg/dL (NR 83-193) and C4 11.5 mg/dL (NR 15-57). Urinalysis, thyroid function test, electrocardiogram and chest x-ray were normal. Echocardiography showed normal mitral leaflets with trace aortic and mitral regurgitation. MRI brain was performed which showed a few T2 flair, high signal punctate foci in bilateral frontal lobes i.e. nonspecific demyelinating foci. Further investigation showed positive anticardiolipin IgG and antibeta 2 glycoprotein IgG antibody in the titres of 418 U/ml and 312U/ml respectively.

After having all the available investigation results, we scored her on the EULAR 2019 classification criteria for SLE.[1]With features of neurological involvement (chorea), leukopenia, low C3, positive anti-dsDNA and antiphospholipid antibodies, the patient scored 14 in the EULAR 2019 classification criteria.\sout[1]The patient was then diagnosed as Neuropsychiatric SLE and was started on hydroxychloroquine, azathioprine and prednisolone.Aspirin was added to her treatment and was planned for repeat antiphospholipid antibody titres after 12 weeks. As the choreiform movements gradually decreased and ceased after the first few days of hospitalization, haloperidol was tapered off.

4 Conclusions/outcome

She is doing well on medications; is currently asymptomatic and has resumed school.

5 Discussion

This case report presents a 12year old girl, whose SLE diagnosis was revealed by a new onset chorea as part of acute neurological manifestation of SLE along with, bicytopenia, elevated titers of anti-dsDNA antibodies, positive antinuclear antibodies, and positive antiphospholipid antibodies.

While chorea can manifest as a symptom in various diseases, when a child exhibits generalized chorea as the sole symptom, Sydenham's chorea is typically the initial diagnosis in regions including Nepal with a high prevalence of Rheumatic Heart Disease.[5]The observation of leukopenia, lymphopenia, and an elevated erythrocyte sedimentation rate (ESR) raised concerns, leading to the search for an alternative diagnosis that could encompass the multisystem manifestations. Hence, the investigations for autoimmune disease were performed, ultimately revealing a diagnosis of Systemic Lupus Erythematosus (SLE).

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SLE is a common diagnosis in the pediatric population.However it mostly presents in the form of lupus nephritis.[6] The American College of Rheumatology (ACR) nomenclature for neuropsychiatry syndromes in SLE includes 12 central nervous system syndromes and 7 peripheral nervous system syndromes.There is a wide variation in the frequency of NPSLE in various studies. Mild cognitive dysfunction and mood disorders are frequently seen as manifestations of NPSLE (6-80%) whereas movement disorders including chorea are extremely rare ($<$ 1% of SLE patients.[7]

Neuropsychiatric(NP) manifestations are a rare presentation of Lupus.In a study carried out in Turkey among 1107 juvenile SLE patients 149 patients had NP involvement (13.5%). The most common NPSLE findings were headache (50.3%), seizure (38.3%), and acute confusional state (33.6%). A study found the prevalence of movement disorders in SLE to be 9.4%.[8]

Chorea as the sole initial presentation of SLE remains rare across all age groups. It manifests in only around 1–3% of cases of SLE.[9]

The exact mechanism behind lupus-associated chorea remains uncertain, but it is potentially linked to damage in the vascular, neuronal, and glial components. There is a high incidence of antiphospholipid antibodies (aPL) in studies of SLE patients with neuropsychiatric manifestations, between 25 and 90%, perhaps implicating a role of these antibodies in the pathogenesis.[10]Vascular injury may involve both inflammatory and thrombotic processes. The presence of aPL appears to have a significant impact on the pathogenesis of neuropsychiatric systemic lupus erythematosus (SLE)[11]

The European League Against Rheumatism (EULAR) recommends antiplatelet therapy for SLE patients with movement disorders and positive aPL antibodies. Anticoagulation agents are only suggested for patients with thrombotic manifestations. As there are no specific guidelines for children with neuropsychiatric SLE, the management should be individualized and based on the decision of the treating clinician.[12]

As chorea is a frequent manifestation of rheumatic fever, the diagnosis of systemic lupus erythematosus (SLE) can be delayed, particularly in regions where rheumatic fever is widespread. Despite its rarity, it is essential to consider the possibility of childhood SLE in the differential diagnosis of any child presenting with unexplained neurological symptoms.

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