

## Psychiatric manifestations of neuro-lupus: A case report

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### Abstract

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that can affect the skin, joints, and internal organs. Among its manifestations, neuropsychiatric involvement, collectively referred to as neuro-lupus, is one of the most severe, including central and peripheral neurological syndromes, autonomic nervous system disorders, and psychiatric syndromes.

We report the case of a 14-year-old girl admitted for acute-onset agitation, auto- and hetero-aggressiveness, and visual and auditory hallucinations. The adolescent was hospitalized in the Neuropediatric and Neurometabolic Diseases Unit at Pediatric Department II of Rabat Children's Hospital, and the diagnosis of neuro-lupus was established based on clinical and biological criteria. Her course was marked by significant clinical improvement under corticosteroid boluses.

The diagnosis of neuro-lupus relies on the exclusion of infectious, metabolic, endocrine, or drug-related causes, and on correlation with SLE immunological activity. Management involves corticosteroids, immunosuppressive agents, and in severe or refractory cases, treatments such as rituximab or plasmapheresis. Despite these therapies, relapses remain frequent, and some patients develop chronic psychotic disorders.

**Keywords:** Systemic Lupus Erythematosus; SLE; Agitation; Psychotic Disorders

### 1. Introduction

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease predominantly affecting the skin and joints. Neurological and psychiatric manifestations are among the most severe visceral involvements and are collectively referred to as neuro-lupus. They include central and peripheral neurological syndromes, autonomic nervous system involvement, and psychiatric syndromes. These manifestations may result from inflammatory or ischemic processes, dysfunction of the blood-brain barrier, or the action of circulating mediators or molecules in the cerebrospinal fluid.

Arguments supporting a link between a neuropsychiatric manifestation and lupus include: the exclusion of differential diagnoses (infectious, metabolic, endocrine, nutritional causes, and primary psychiatric disorders or those secondary to medication or toxins), the inaugural or early occurrence of neuropsychiatric manifestations (within the first year after lupus diagnosis in 50–60% of cases), and the clinical and immunological activity of the lupus disease.

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## 2. Case Report

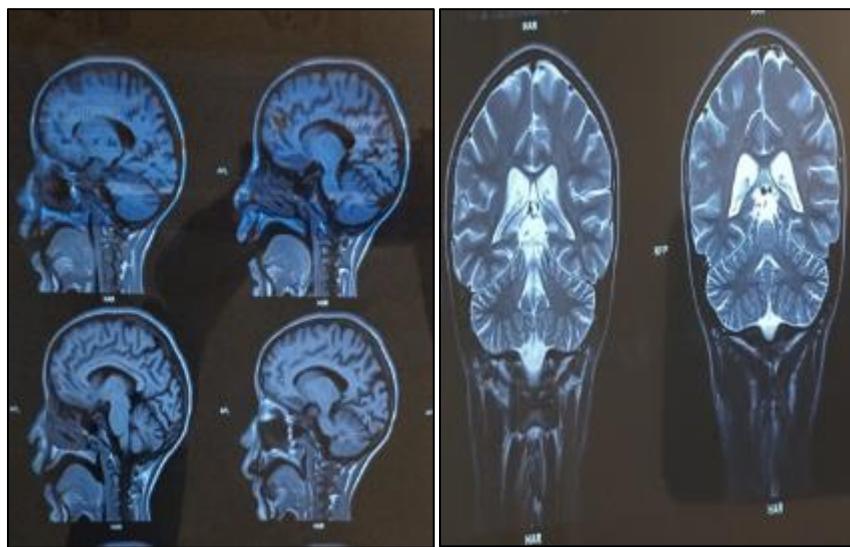
This is a 14-year-old adolescent girl with a medical history of inflammatory arthralgia, hypertension treated with beta-blockers since age 12, sensorineural hearing loss with decreased visual acuity, second-degree consanguinity, and a family history of intellectual disability and hearing loss in cousins.

The patient had been presenting for 2 months with behavioral disturbances, including auto- and hetero-aggressiveness, delusions, and visual and auditory hallucinations. Clinical examination revealed a highly agitated child with a fleeting gaze. Vital signs were within normal limits. The rest of the physical examination was unremarkable, except for a decrease in segmental muscle strength in both lower limbs symmetrically, with low-intensity deep tendon reflexes.

Laboratory tests showed an elevated ESR at 33 mm, negative CRP, and a doubtful Coombs test. Toxin screening was negative. Cerebrospinal fluid analysis was normal. Urinalysis revealed aseptic leukocyturia. Viral serologies were negative. Antinuclear antibodies (ANA) were positive, and anti-dsDNA antibodies were positive. Antiphospholipid antibodies were negative. Thyroid function tests were normal, and anti-TPO antibodies were negative.

Cerebro-medullary MRI showed diffuse widening of cortical sulci both supra- and infratentorial. MR spectroscopy was unremarkable. EEG revealed a pattern of global cerebral distress without epileptiform activity. Electromyography showed a length-dependent sensorimotor polyneuropathy. Echocardiography revealed moderate left ventricular hypertrophy. Visual evoked potentials demonstrated bilateral cortical response impairment consistent with PR chiasmatic involvement. Renal function was normal, and 24-hour proteinuria was negative.

The patient was treated with monthly corticosteroid boluses for 6 months (30 mg/kg/day for 3 days), intravenous immunoglobulins (1 g/kg/day for 2 days), and psychiatric management (anxiolytics and antipsychotics). After 2 corticosteroid boluses, there was a marked regression of auto- and hetero-aggressiveness, a decrease in delusions and hallucinations, and negotiation of antibodies. Follow-up MRI showed persistence of diffuse widening of cortical sulci both supra- and infratentorial.



**Figure 1 and 2** Brain MRI of the patient showing widening of the cortical sulci both supra- and infratentorial

## 3. Discussion

Neuropsychiatric manifestations of systemic lupus erythematosus (SLE) in children represent a major diagnostic and therapeutic challenge. They occur in approximately 20–40% of pediatric cases and may be inaugural in nearly one-third of cases [1]. Clinical presentation is highly variable, including central and peripheral neurological involvement, or isolated psychiatric disorders such as psychosis, anxiety, or behavioral disturbances [2]. In our patient, the initial presentation was dominated by severe psychiatric symptoms, illustrating the complexity of early recognition of pediatric neuro-lupus.

Diagnosis primarily relies on the combination of clinical, biological, and radiological findings, while carefully excluding infectious, metabolic, or drug-related differential diagnoses [3]. Antinuclear antibodies (ANA) and anti-dsDNA antibodies are frequently positive, reflecting autoimmune activity. Decreased complement fractions (C3, C4) also serve as markers of lupus flares [1,3]. However, the correlation between laboratory findings and neuropsychiatric manifestations remains imperfect, justifying a multidisciplinary approach involving pediatric neurologists, psychiatrists, and rheumatologists [4].

Pathophysiologic ally, central nervous system involvement in neuro-lupus may result from direct inflammation, ischemic microangiopathy, blood-brain barrier disruption, or the effects of cytokines and neurotoxic autoantibodies [2,4]. This diversity explains the variability in clinical and radiological profiles, sometimes rendering brain imaging normal despite severe symptoms, as observed in our case.

Treatment of neuro-lupus classically relies on high-dose corticosteroids, often combined with immunosuppressive agents such as cyclophosphamide in severe cases [2,3]. Intravenous immunoglobulins may be used as adjunct therapy, particularly in cases of contraindication or resistance to cytotoxic agents. In refractory cases, biologics such as rituximab have shown significant clinical benefit, especially in children [5]. Early use of these targeted therapies appears to improve functional prognosis and reduce relapses.

Despite therapeutic advances, pediatric neuro-lupus remains associated with a risk of chronic neurological and psychiatric sequelae, warranting long-term follow-up. Regular neuropsychological evaluation and integrated psychiatric management are essential to optimize quality of life and school reintegration.

Management of pediatric neuro-lupus requires a multidisciplinary approach, mainly based on corticosteroid boluses and sometimes other immunosuppressive therapies. Despite treatment, neuro-lupus carries a poor prognosis, particularly due to its frequent association with renal involvement. Early treatment is essential to limit morbidity and mortality.

Our patient responded well to six monthly methylprednisolone boluses (3-day courses each month); her psychotic symptoms regressed, although sensorimotor deficits in the lower limbs persist.

#### 4. Conclusion

Pediatric neuro-lupus remains associated with a risk of chronic neurological and psychiatric sequelae, necessitating prolonged follow-up. Regular neuropsychological assessment and integrated psychiatric care are essential to optimize quality of life and school reintegration.

#### Compliance with ethical standards

##### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

##### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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