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Neuro lupus Revealed by an Acute Psychiatric Episode in a 14 Year Old Adolescent Girl: A Case Report

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Abstract **Case Report**

Introduction: Neuro-lupus, the neuropsychiatric form of systemic lupus erythematosus (SLE), may present in an atypical even inaugural manner, particularly in children and adolescents. Although relatively rare compared with the adult form, juvenile lupus often exhibits increased severity and more rapidly progressive clinical manifestations, including its neuropsychiatric component. The objective of this work is to emphasize the importance of suspecting, identifying, and managing neuro-lupus early in pediatric patients presenting with unexplained mental disorders, in order to prevent any diagnostic delay that could affect prognosis. ***Case presentation:*** We report the case of a 14-year-old Moroccan adolescent girl, Hasna B., admitted to the pediatric emergency department for psychomotor agitation. Her early development was globally delayed, and for one year she had exhibited progressive social withdrawal, mistrust toward her family, and verbal hallucinations, all of which led to discontinuation of schooling. Despite this deterioration, no management had been instituted. The psychiatric examination objectified a catatonic syndrome associated with a hallucinatory syndrome, while her history notably included inflammatory arthralgias, an untreated femoral fracture, arterial hypertension of indeterminate etiology, and a family history of deafness. Additional investigations demonstrated positive antinuclear antibodies, cerebellar atrophy on magnetic resonance imaging, global cerebral dysfunction on electroencephalogram, a sensorimotor polyneuropathy on electroneuromyogram, and bilateral papilledema on ophthalmologic examination. In view of these results and the clinical picture, the diagnosis of juvenile neuro-lupus was retained. ***Discussion:*** Juvenile lupus may account for up to twenty percent of all lupus cases. Compared with adults, it is characterized by often more severe involvement, a female predominance, and an increased risk of neurologic or psychiatric involvement (neuro-lupus). Associated neuropsychiatric manifestations (headaches, mood disorders, psychoses, epilepsy, etc.) may affect up to fifty-six percent of children and adolescents and justify heightened vigilance by clinicians. When these psychiatric disorders are directly related to lupus activity, management is based mainly on corticosteroid therapy and immunosuppressants, while psychotropic drugs are used as complementary and temporary measures. In this context, it is also important to monitor organ functions, notably renal function, to prevent any medication overdose. ***Conclusion:*** This observation clearly illustrates the importance of investigating an organic etiology—particularly neuro-lupus—before any atypical psychiatric presentation or one resistant to usual treatments in children and adolescents. Early identification of juvenile lupus allowing targeted treatment (corticosteroid therapy, immunosuppressants) can profoundly modify the clinical course and prognosis of the disease, underscoring the need for a multidisciplinary approach and constant vigilance in the evaluation of pediatric neuropsychiatric disorders.

Keywords: neuropsychiatry, neuro-lupus, systemic lupus erythematosus, child and adolescent psychiatry.

INTRODUCTION

Nowadays, it is widely recognized that numerous organic pathologies may manifest through psychiatric symptoms, sometimes inaugural, even before the appearance of so-called "classic" clinical signs. This observation echoes the common history of neurology and psychiatry, two disciplines once closely linked (under the term "neuropsychiatry") before gradually separating. The rise of psychoanalysis and the desire to distinguish "organic" from "functional" disorders contributed to this split. However, advances in neurosciences and brain imaging today tend to re-establish a bridge between these two specialties by highlighting the biological bases underlying many psychiatric disorders. In this context, the management of a patient presenting with psychiatric symptoms must always include the possibility of an underlying neurologic or systemic pathology, the

identification of which can radically change prognosis. This is notably the case for systemic lupus erythematosus (SLE), an autoimmune disease that preferentially affects the skin and joints but whose neurologic involvement (neuro-lupus) may manifest as psychiatric presentations. Juvenile lupus, even rarer, represents approximately 10 to 20% of lupus cases and may present at an early age, often with more severe clinical forms. We present the case of a 14-year-old adolescent girl in whom the diagnosis of juvenile neuro-lupus was retained only after an insidious evolution of psychiatric symptomatology, which then abruptly worsened in the form of psychomotor agitation with a clastic crisis, thus highlighting the need to investigate potential organic causes in pediatric patients presenting with mental disorders.

Case Presentation

H. B. is a 14-year-old Moroccan adolescent girl, out of school for one year, initially admitted to the pediatric emergency department for psychomotor agitation. Her father is a shopkeeper, her mother a housewife, and she is the product of a second-degree consanguineous marriage. She has a healthy 17-year-old older sister. Regarding development, pregnancy and delivery were reportedly uneventful, but there was a global delay in her early development: head control at 7 months, sitting at 9 months, walking at 24 months, first words at 24 months and first sentences at 36 months, acquisition of daytime and nighttime bladder control around age 5. Her school performance was always poor and progressed only thanks to teachers' indulgence. Relationally, she was described as sociable and extroverted before the onset of symptoms. For about one year, her behavior progressively changed: she isolated herself, spoke less and less with her family, then developed marked mistrust toward those around her, particularly toward her mother. Her relatives caught her several times talking to herself. Despite the negative impact on her functioning, which notably resulted in school discontinuation, no management was considered. It was only after a psychomotor agitation crisis, marked by a destructive violent outburst, that she was finally brought to pediatric emergency care. During hospitalization in the pediatric department, Hasna remained difficult to manage due to agitation, food refusal, insomnia, and lack of interaction with the environment. She initially received 0.5 mg/day of risperidone without notable improvement after two days, which led to requesting a child psychiatry consultation and additional management. Her medical history included intermittent inflammatory arthralgias evolving over two years (not followed), arterial hypertension of indeterminate etiology (on beta-blocker), an untreated fracture of the distal third of the right femur, and scoliosis; as well as a family history of deafness and intellectual developmental disorders in cousins. Psychiatric examination revealed catalepsy, waxy flexibility, negativism, mutism, and occasional listening postures, all suggesting a catatonic syndrome associated with a hallucinatory syndrome. The remainder of the clinical examination revealed difficulty walking due to the fracture, lower-limb amyotrophy with absence of deep tendon reflexes, while muscle strength remained preserved in the upper limbs. In light of this clinical symptomatology, several diagnoses were considered, including an organic cause potentially explaining this psychiatric presentation, catatonia due to another medical condition, and early-onset schizophrenia. Complementary investigations showed, notably, positive antinuclear antibodies (ANA), cerebellar atrophy on brain and spinal magnetic resonance imaging (MRI), an electroencephalogram indicating global cerebral dysfunction without epileptic activity, an electroneuromyogram suggesting a sensorimotor polyneuropathy, bilateral papilledema on ophthalmologic examination, and bilateral sensorineural hearing loss on audiogram. Following these examinations and in view of the clinical picture, the diagnosis of neuro-lupus was retained.

Discussion

We report here the observation of a 14-year-old adolescent girl who's insidiously evolving psychiatric symptoms ultimately revealed juvenile neuro-lupus. To better understand the scope of this diagnosis, certain aspects of systemic lupus erythematosus should be recalled: the most frequent diagnostic age range lies between 15 and 44 years, but it may also occur in children and adolescents under the name of juvenile lupus, before age 18. Although it shares numerous clinical and biological characteristics with the adult form, it presents certain specificities, notably potentially increased severity, more rapidly progressive symptoms, and sometimes more complex management. Relatively rare, juvenile lupus represents approximately 10 to 20% of all cases, with a median diagnostic age between 10 and 15 years. As in adults, a female predominance is noted, with a female-to-male ratio possibly reaching 4 or 5 to 1. Beyond these epidemiological and clinical particularities, juvenile lupus is characterized by more frequent renal involvement and a higher risk of neurologic or psychiatric complications, often grouped under the term "neuro-lupus." These manifestations may affect up to fifty-six percent of affected children and adolescents, underscoring the need for vigilance and a multidisciplinary approach in this population.

The most frequent symptoms are headaches (twenty-eight percent), mood disorders (twenty-one percent), neurocognitive disorders (twenty percent), epilepsy (ten percent), and anxiety disorders (six percent). Imaging by magnetic resonance may reveal cerebral atrophy, white- or gray-matter abnormalities, as well as lacunar lesions. Lumbar puncture is often performed to exclude an infectious cause. When psychiatric symptoms are directly linked to lupus involvement, management relies essentially on immunosuppressive therapy or corticosteroid therapy, while the use of psychotropic medications remains

temporary and adapted to symptomatology. Precautions include avoiding certain drug classes (phenothiazines, lithium, carbamazepine) and monitoring organ functions, particularly renal function, to prevent overdose risk. In addition to neurologic or psychiatric manifestations, other forms of involvement are frequently observed in children: cutaneous (malar rash, photosensitivity, alopecia), articular (arthralgias, arthritis), renal (lupus nephritis, proteinuria, hematuria, renal failure), hematologic (hemolytic anemia, leukopenia, thrombocytopenia), and serologic (antinuclear antibodies, anti–double-stranded deoxyribonucleic acid antibodies, antiphospholipid antibodies). This clinical diversity illustrates the complexity of lupus in children and underscores the need for a multidisciplinary approach. Among the most frequent neuropsychiatric manifestations of neuro-lupus are first mood disorders, whose prevalence ranges between six and twelve percent. Individuals with lupus appear more prone to developing these disorders than the general population, and depressive episodes may be particularly marked during disease flares. It nevertheless remains difficult to determine whether this is a purely reactive phenomenon or directly linked to lupus activity. Bipolar disorder is clearly rarer. In the absence of specific recommendations for mood disorders associated with lupus, monitoring therapeutic adherence (immunosuppressants, other treatments) in depressed patients is crucial. In severe depressive forms, if a link with lupus disease is likely or in the presence of signs of disease activity, initiation (or intensification) of immunosuppressive treatment may be considered in addition to antidepressant therapy. Notably, corticosteroid therapy does not appear to increase suicide risk. Anxiety and obsessive-compulsive disorders, whose prevalence is estimated between one and twenty-four percent, encompass generalized anxiety, panic disorder, and various forms of phobias (agoraphobia, social phobia, specific phobias). Contributing factors to anxiety include difficulties adapting to the stress of a chronic disease, but also cutaneous lesions likely to have aesthetic repercussions and promote agoraphobia or social phobia. No specific treatment is validated for lupus-related anxiety, but cognitive-behavioral therapies and group therapies—already recommended for anxiety disorders—may prove useful. Pharmacologic treatment may be instituted if necessary. Psychosis constitutes another important facet of these manifestations and may inaugurate the disease in twenty-one to fifty-four percent of cases. Most psychotic patients also present other clinical signs (cutaneous, articular, renal involvement, etc.). When psychosis is attributed to lupus, the combination of corticosteroid therapy and immunosuppressants leads to improvement in sixty to eighty percent of cases, generally within two to four weeks. Antipsychotic treatment may be indispensable, and relapses remain possible. Approximately twenty percent of patients may retain moderate long-term psychotic symptoms. The forms of delusion (paranoid, visual or auditory hallucinations, grandiose delusion) vary among studies; magnetic resonance imaging often remains normal (sixty to seventy percent of cases), and cerebrospinal fluid may also be normal or show only discrete abnormalities (moderate pleocytosis or elevated protein). Delirium, rarer in adult lupus, nevertheless affects up to thirty-five percent of cases in certain pediatric studies. It most often signals an underlying metabolic, infectious, or vascular cause. In these situations, magnetic resonance imaging, lumbar puncture, and electroencephalogram are indispensable to guide diagnosis.

Corticosteroid-induced psychiatric disorders mainly include anxious, depressive, or psychotic manifestations, although manic or hypomanic episodes may occur, especially during the first month of high-dose treatment (approximately one milligram per kilogram per day). These phenomena are generally associated with doses exceeding forty milligrams of corticosteroids per day and may be favored by hypoalbuminemia or hypocomplementemia. Evolution is most often favorable once corticosteroid therapy is reduced; if symptoms persist, the hypothesis of direct imputability to the corticosteroid should be re-examined. In any case, it is recommended not to reintroduce the same dose upon resumption of treatment, and psychiatric support may also be necessary.

Catatonia, although described in only about twenty cases in lupus patients, appears to respond to high doses of benzodiazepines. Headaches, affecting ten to twenty percent of individuals with lupus, may present in various forms ranging from migraine to intracranial hypertension to cluster headaches. Treatment varies according to the cause: simple symptomatic management for a “benign” headache, anticoagulants in the event of cerebral thrombophlebitis, blood pressure and renal function control in reversible posterior encephalopathy, or finally corticosteroid therapy and immunosuppressants in lupus flare with severe signs (intracranial hypertension, altered vigilance).

Finally, epilepsy may take all forms encountered in the general population, whether generalized seizures (tonic-clonic, atonic, absences, myoclonic) or focal (simple or complex). If seizure activity precedes the diagnosis of lupus, drug-induced lupus from anticonvulsants should be considered. Moreover, an estimated three to fifty-one percent of children with lupus will present at least one convulsive seizure, whereas the prevalence of epilepsy in the general pediatric population remains clearly below one percent.

CONCLUSION

In the face of a psychiatric presentation—particularly in children—several clinical elements should raise suspicion of an organic origin. These may include an unusual or early onset of symptoms, brief episodes with spontaneous fluctuations, resistance to usual treatments, neurologic signs (abnormal movements, gait disorders, confusion), or involvement

of multiple organs. Likewise, the presence of an inflammatory syndrome or organ dysfunction on biological testing constitutes an additional warning sign.

Beyond neuro-lupus, various neurologic pathologies may manifest similarly, notably autoimmune encephalitides (such as multiple sclerosis, systemic lupus erythematosus, or anti–N-methyl-D-aspartate receptor antibody encephalitis), infectious encephalitides (herpetic, human immunodeficiency virus infection, cerebral toxoplasmosis, Lyme disease), temporal lobe epilepsy, remote cranial trauma, frontal or temporal brain tumors, hydrocephalus, cerebral malformations, idiopathic cerebral calcifications, as well as certain pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections or pediatric acute-onset neuropsychiatric syndromes.

Early identification of an underlying organic pathology is essential, as specific diagnosis and treatment can profoundly modify the clinical course and, in some cases, prove decisive for recovery. This reality demands a multidisciplinary approach and constant vigilance in the face of any atypical psychiatric presentation in children and adolescents.

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