

**FEBRILE CONVULSIONS REVEALING SYSTEMIC LUPUS ERYTHEMATOSUS IN CHILDREN OBSERVED AT THE CHU ANDRAINJATO FIANARANTSOA****Ramamonjirinirina Tahina Prudence\*<sup>1</sup>, Rabemananjara Antsa<sup>2</sup>, Rakotomahefa Mbola<sup>3</sup> and Rabesandratana Noro<sup>4</sup>**<sup>1</sup>Department of Pediatrics CHU Andrainjato Fianarantsoa.<sup>2</sup>Pediatric Service CHU Tambohobe Fianarantsoa.<sup>3</sup>Department of Pediatric Oncology.<sup>4</sup>Neonatology Department, Mother and Child Complex, Chu Pzga Majunga.**\*Corresponding Author: Ramamonjirinirina Tahina Prudence**

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**ABSTRACT**

**Introduction:** Pediatric systemic lupus erythematosus is a rare condition. Our aim was to report a case of systemic lupus erythematosus revealed by seizures in a 9-year-old girl. **Observation:** We report a case of 9-year-old girl hospitalised in the paediatric service of University Teaching Hospital of Andrainjato Fianarantsoa for generalised tonic-clonic seizures. The diagnosis of SLE was based on a butterfly-shaped malar rash, oral ulceration, seizures, anaemia, and positive anti-native DNA and antinuclear antibodies. Corticosteroid therapy combined with a synthetic antimalarial drug led to an improvement in the cutaneous signs and a progressive regression of the articular signs. **Conclusion:** Seizure may be the mode of manifestation of SLE in children. The presence of skin and joint signs can easily guide the diagnosis.

**KEYWORDS:** Child, seizures, systemic lupus erythematosus.**INTRODUCTION**

Systemic lupus erythematosus (SLE) is rare in paediatric population. It is a condition that is often diagnosed before the age of 16 in 10-15% of cases.<sup>[1,2]</sup> It is often considered more severe than in adults because of the high frequency of severe kidney, haematological and neurological damage.<sup>[3]</sup> We report a case of a 9-year-old girl presenting a seizure revealing a systemic lupus erythematosus.

**Case Presentation**

A 9 year old Malagasy girl was referred to our department for a generalized convulsive seizure. Since 4 months, she had been complaining of recurrent arthralgia of the knees since 3 to 4 weeks prior to her presentation. followed, by the occurrence of bilateral erythematous photosensitive lesions in the malar region of the face. No diagnosis of lupus had been made during the medical consultations. Then, she was treated with antalgic.

The day before her admission, she presented a generalised tonic-clonic seizure lasting for about 4 minutes with non precised fever without postcritical alteration of consciousness.

The child was born at full term and well vaccinated. She had no medical and familial history; Upon arrival, she had a high temperature (38.2°C), with a heart rate of 130 per minute, a blood pressure of 100/60 and a respiratory rate of 24 breaths per minute. She was conscious with a Glasgow score of 15/15, but asthenic. She complained of joint pain in the left and right knees scored at 5/10 on the VAS (Visual Analogue Scale). The neurological finding was normal while the skin examination revealed bilateral erythematous eruptions on the malar region of the face with oral ulceration (figure 1) and bilateral erythema on the plantar surfaces (figure 2).

**Figure 1: Malar Butterfly rash and oral ulceration.**



**Figure 2: Plantar erythema.**

The rapid diagnostic test for malaria was negative. Urine analysis showed 2+ proteinuria and there was no urinary tract infection biomarker.

These are the results of laboratory test: microcytic anaemia with a haemoglobin level (8 g/dl); leukocytes (7400 G/l) and lymphocytes (1920 G/l); sedimentation rate (60 mm); ferritin level (1613 µg/l); C-Reactive Protein (6 mg/L); Creatinine (normal); Transaminase (double of normal), and lactate dehydrogenase LDH (3 times normal); Emmel's test (negative); anti-native DNA antibody test was positive (20 IU/mL); the anti-nuclear antibody test (1280 IU/mL).

X-rays of the knees did not reveal any particular abnormality. Computed Tomography of the brain, brain magnetic resonance imaging and electroencephalogram could not be performed.

In view of the presence of 5 out of 11 ACR criteria in 1997,<sup>[4]</sup> such as butterfly wing malar rash, oral ulceration, seizures, positive anti-native DNA and antinuclear antibodies, the diagnosis of SLE was retained. Face to the neurological involvement and the positive proteinuria at 2+, a corticosteroid therapy using Prednisone at a dose of 2 mg/kg/d associated with a synthetic anti-malarial using hydroxychloroquine at a dose of 8 mg/kg/d had been initiated.

An improvement of skin signs was noted from the first week of treatment with progressive regression of joint signs. Biological markers of inflammation were progressively normalised and the 24-hour proteinuria test carried out 48 hours after the start of corticosteroid therapy b negative.

The hospitalization period lasted for 5 days and a regular weekly and then monthly follow-up was planned after the hospitalization to look for signs of a possible relapse, to monitor the side effects of the drugs (blood pressure, heart rate, infectious outbreaks), and to follow the growth.

## DISCUSSION

A case of SLE fulfilling 5 out of 11 ACR criteria revealed by seizures in a 9-year-old girl was reported. Neurological investigations, such as EEG and brain CT scan could not be performed due to the lack of funds.

In the United States, the prevalence of SLE is 15 to 50 per 100,000 population.<sup>[5]</sup> It is a rare condition in children under 16 year-old with a prevalence of 0.5 per 100,000.<sup>[5]</sup> The predominance of females in systemic lupus erythematosus may be explained by the role of hormonal factors.<sup>[6]</sup> and increases with age.

The clinical presentation in children is similar in adult population. The clinical manifestations are polymorphous, sometimes deceptive at first, thus delaying the diagnosis in the absence of specific cutaneous signs,<sup>[7]</sup> as was the case in our patient. The diagnosis is easier in the presence of initial specific skin involvement such as butterfly wing malar rash (27%) and cutaneous vasculitis (45%). In children, joint involvement (96%) and haematological involvement are also the most common.<sup>[8]</sup>

The patient's 24-hour proteinuria test was negative but performed 48 hours after the start of steroid therapy, while the analyse of urine on admission was positive with two crosses of proteinuria. This proteinuria that disappeared after 48 hours of corticosteroid therapy could not be considered as persistent proteinuria. The creatinine level was normal. It was difficult to include this proteinuria among the criteria for the diagnosis of SLE. Renal involvement is present in 30-80% of paediatric lupus cases and is a serious factor.<sup>[9]</sup> Renal biopsy is essential in this case but was not performed in our case due to the rapid resolution of proteinuria (48hours after corticosteroid therapy), although it is possible to perform at our centre.

Seizures are rare. They occur in 7-10% of all patients with SLE and 23.4% those with neurolupus.<sup>[10]</sup> In these patients, brain MRI does not show any particular abnormality in 27- 60% of cases. In this situation, seizures are said to be "cryptogenic" or of undetermined etiology and often of single episode, as in our patient's case. Nevertheless, possible causes of seizures such as neurological infections, electrolyte disorders, brain tumours with no direct links to SLE, or neuropsychiatric syndromes in favour of neurolupus, or iatrogenic causes.<sup>[11]</sup> must first be eliminated, the latter having been previously excluded in our patient's case. Thus it was retained a neurological attack of lupus.

Treatment depends on the organs affected. In our case, neurological involvement associated with a suspicion of renal involvement in front of proteinuria (not confirmed as persistent) were considered as criteria of severity of lupus. Thus, corticosteroid therapy was started in parallel with antimalarial treatment. The management and prevention of relapses is based on the same

recommendations as in adults,<sup>[10]</sup> Corticosteroid therapy is often necessary in children and should be used in low doses in mild forms, and early in high doses in severe forms. Hydroxychloroquine should be prescribed systematically. The latest recommendations of the French group for the study of SLE in children propose corticosteroids in severe and renal forms, and synthetic antimalarials to avoid relapses,<sup>[12]</sup> Antiepileptic drugs are indicated in cases of recurrent or partial seizures, neurological deficits, structural abnormalities on brain imaging or EEG abnormalities.<sup>[10]</sup>

## CONCLUSION

Seizures may be the mode of revelation of SLE in children. It is indispensable to look for other, manifestations that are often discreet, as their presence can easily guide the diagnosis. Early diagnosis for adequate management is necessary to reduce its morbidity and mortality.

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