SAS Journal of Medicine

Abbreviated Key Title: SAS J Med ISSN 2454-5112 Journal homepage: <u>https://saspublishers.com</u> **∂** OPEN ACCESS

Rheumatology

Juvenile Systemic Lupus about an Observation and Review of the Literature

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DOI: 10.36347/sasjm.2022.v08i09.002

| **Received:** 13.08.2022 | **Accepted:** 07.09.2022 | **Published:** 14.09.2022

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Abstract

Case Report

Introduction: Systemic lupus erythematosus (SLE) is a non- organ specific auto-immune disease of multifactorial origin (genetic, immunological, and environmental) acting in concert. The one with juvenile-onset is rare, renal involvement is the most frequent complication requiring mostly corticosteroid treatment. *The objective of the Work:* To show the importance of early diagnosis and management of juvenile systemic lupus with insidious evolution for a good functional and vital prognosis. *Clinical Case:* It was a 13-year-old girl carrying sickle cell trait (SCT) as well as her two parents, referred from pediatrics who present for 3 years chronic peripheral dry polyarthritis associating photosensitivity, biological analysis finds hemolytic anemia (hemoglobin level at 7.3g/dl), an inflammatory syndrome (VS at 98mn at the 1st hour and CRP at 56mg/l), transaminases, uremia, creatinemia, and glycemia were normal, but rheumatoid factors, antinuclear antibodies 1/210 type speckled aspect, U1RNP, anti-Sm and anti-SSA/Ro 52 came back positive following the new criteria of the SLICC group (Systemic Lupus International Collaborating Clinics) the diagnosis of juvenile lupus is retained, then put under treatment based on Prednisone 5 mg/d, Hydroxychloroquine 200 mg /d, blood transfusion (350ml iso rhesus group) The evolution was favorable with SLEDAI at 3.17 at the 3rd month and 2 at the 6th month. *Conclusion:* Juvenile lupus is more severe than adult lupus in terms of renal complications, corticosteroid treatment is a necessity which is not without side effects on growth.

Keywords: Juvenile systemic lupus Senegal.

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a nonorgan specific auto-immune disease of multifactorial origin (genetic, immunological, and environmental) acting in concert, first described by Hippocrate, his first description was published by Laurent Theodore Biett and his student Pierre Louis Alphee Cazenave in 1833 [1]. Juvenile onset is rare, with 15% of cases beginning before the age of 16 [2, 3]. Renal, hematological and neurological involvement are the most frequent complications in children compared to adults, requiring most often corticosteroid therapy [4].

We report a case of a 13-year-old girl with a review of the literature.

This was a 13-year-old girl with sickle cell anemia (AS) and both of her parents, who presented for 3 years with bilateral, symmetrical, additive, and persistent chronic dry polyarthritis of the wrists, elbows and knees associated with photosensitivity. The biological analysis found hemolytic anemia (hemoglobin level 7.3g/dl), an inflammatory syndrome (SV 98mn at the 1st hour and CRP 56mg/l), transaminases, uremia The transaminases, uremia, creatinemia, and glycemia were normal, but the rheumatoid factors, the anti-nuclear antibodies (1:210), U1RNP, anti-Sm and anti-SSA/Ro 52 were positive, of wrists, elbows, knees, and lung did not show any particularities, so the diagnosis of systemic lupus is retained in front of the non-erosive peripheral polyarthritis, the presence of antibodies (antinuclear, U1RNP, anti-Sm, anti-SSA/Ro 52) by the new criteria

OBSERVATION

Citation: Adama BAH, Ramadhane BOUCHRANE, Harine Abdel Aziz GARBA, Moussa TRAORE, Mamadou Diouldé KANTE, Aïssatou N'diaye, Chekh Ahamad Bamba DIAO, Moustapha NIASSE, Saïdou DIALLO. Juvenile Systemic Lupus about an Observation and Review of the Literature. SAS J Med, 2022 Sept 8(9): 594-596.

of the SLICC group (Systemic Lupus International Collaborating Clinics) she received treatment based on Prednisone 5 mg/d, Hydroxychloroquine 200 mg/d, blood transfusion (350ml of whole blood iso rhesus group). The evolution was favorable with SLEDAI at 3.17 in the 3rd month and 2 in the 6th month.

DISCUSSION

Our observation is one of juvenile systemic lupus which is of epidemiological, diagnostic, prognostic and therapeutic interest.

Systemic lupus in children is a rare, polymorphic disease that is more severe than the adult form and most often requires immunosuppressive therapy because of the greater frequency of renal involvement [5, 6].

The prevalence of systemic lupus in children is much lower than in adults. Its annual incidence in children under 16 years of age is less than 1 per 100,000 people in studies from Europe and North America. In Taiwan, its prevalence has been estimated at 6.3 per 100,000 [7].

It differs from that of adults by a lower sex ratio of 1:5 to 1:18 [8], as in our case, which is female.

The nonspecific symptoms are fever, anorexia, weight loss, and asthenia. They are particularly misleading in adolescence. Early in the course of the disease, a single organ may be affected, but the systemic form is the usual form of disclosure. Arthritis, rash, and renal involvement are the most common involvement in the pediatric form [9]. Our teenager presented with bilateral, symmetrical, additive and persistent chronic dry polyarthritis involving the wrists, elbows and knees, these signs are consistent with the literature data that arthralgia or arthritis of small and large joints affects nearly 80% of cases [9].

According to Cameron, 75% of children with lupus will develop lupus nephropathy and almost half of them have WHO stage IV renal disease at diagnosis [3, 10]. The goal of treatment is to achieve remission of the disease by requiring an adapted treatment to limit side effects, in particular on growth and development. Photoprotection is essential and relies on the application of sunscreen. Vaccination should be as broad as possible, taking into account the type of vaccine and current treatments, and including vaccination against pneumococcus and meningococcus. Treatment regimens are often based on those of adults. Corticosteroid therapy plays a central role in the treatment of lupus in children [11] and remains the most widely used treatment according to most authors [11, 12]. It is systematically used by some, in a nuanced way by others, that means in moderate forms uncontrolled by NSAIDs and PSA or in severe visceral forms [13], generally, it is used in high doses (1-2 mg/kg/d) [14].

The prognosis of lupus remains poor, depending on its location. Indeed, the poor prognosis remains linked to renal involvement or, more rarely, to the involvement of the central nervous system or the heart [13, 15].

CONCLUSION

Juvenile systemic lupus is a rare connective tissue disease that impacts growth through corticosteroid therapy for renal involvement.

Early diagnosis and treatment improve the vital and functional prognosis.

Multidisciplinary management allows good statural development.

Sincere Thanks

To all those who participated in the care of the patient in one way or another with a special mention to all the staff of the rheumatology department of the Dantec hospital in Dakar.

DECLARATION OF INTEREST

No interest.

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