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Research Paper

Childhood-onset systemic lupus erythematosus: about 3 cases in sub-Saharan black Africa

AYOUBA TINNI Ismael [1]*, YAMEOGOWendyamNadège[1], OUEDRAOGO Mariam [1], ZABSONRE/TIENDREBEOGO Wendlassida Joelle Stéphanie [1,2], KABORE Fulgence[1,2], BONKOUNGOU Marcellin[1], OUEDRAOGO Dieu-Donné [1,2]

1: Department of Rheumatology of Bogodogo University Hospital
2: University of Joseph Ki-ZERBO of Ouagadougou
** Corresponding author: AYOUBA TINNI Ismael*
Rheumatology Department of Bogodogo University Hospital, 14 BP 371 OUAGADOUGOU 14 Burkina Faso.

Abstract:*Systemic lupus erythematosus (SLE) is rarely reported in children. We report 3 cases of pediatric systemic lupus erythematosus diagnosed in the rheumatology department of the Bogodogo University Hospital, Burkina Faso, after 14 years of rheumatological practice.*
Case report 1: A 9-year-oldschoolgirl received to the rheumatology departmentwithpolyarthritis. The immunological assessment was positive with anti-nucleosome antibodies, anti-native DNA antibodies and anti-ENA antibodies. The evolution was favorable under corticotherapy and hydroxychloroquine.
Case report 2: A 14-year-oldschoolgirl received for diffuse pain and skin lesions located on the uncovered areas. Immunologically, anti-nucleosome, anti-native DNA, anti-histone and anti-ENA antibodies were positive. The evolution was favorable under corticotherapy and hydroxychloroquine.
Case report 3:A 11-year-old first twin,schoolgirl, HIV positive type 1. She was admitted because of prolonged, fever, dyspnea, polyseritis and generalised pruritus. The immunological tests were positive for anti-nucleic acid, anti-native DNA and anti-ENA antibodies. The evolution was favorable under corticosteroids and hydroxychloroquinefor about 6 months.
Conclusion:Pediatric SLE is more severe than the adult form and should be recognised and treated early. Morbidity and mortality depend on the organs affected.
KeyWords:*Systemic lupus erythematosus, Childhood-onset, pediatrics, Burkina Faso, Subsaharian Africa*

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I. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune, multisystemic inflammatory disease of unknown cause, characterised biologically by the production of multiple autoantibodies, the most characteristic of which are directed against certain components of the nucleus such as native deoxyribonucleic acid (DNA) and nucleosomes [1]. It is rare in children, being diagnosed before the age of 16 years in 10-15% of cases, and affecting mainly peripubertal girls. The initial manifestations are varied and sometimes misleading, and severe forms, especially those involving the kidneys, are more common than in adults [2]. In severe forms, the long-term prognosis depends in part on the earlyness and nature of the treatment, which is based on multidisciplinary managementthat takes into account the child's development and growth [3]. We report 3 cases of pediatric lupus diagnosed at the Rheumatology Department of the University Hospital of Bogodogo, Burkina Faso.

II. Case Report

Observation 1:

A 9-year-old schoolgirl had been followed and treated since the age of 5.5 months for epilepsy, with no particular family or pregnancy history. Delivery was by caesarean section for labour dystocia. She was seen in a rheumatology consultation for polyarthritisof 1 year's duration affecting small and large joints bilaterally, symmetrically fixed and additive (IPD, IPP, MCP, wrists, elbows, knees, ankles), in a context of anorexia

without fever, weight loss or asthenia. There were no headaches, conjunctivitis, dry eyes or mouth, nasal or oral ulcers, photosensitivity, Raynaud's disease, alopecia or hematuria. The blood count, liver and kidney function tests were normal. The immunological examination showed anti-nucleosome antibodies >1280, anti-native DNA antibodies at 278 IU/ml, anti-ENA antibodies weakly positive with U1RNP at 10 IU/ml, SSA/Ro, SSB/La, Sm antibodies negative, and anti-histone antibodies. Antiphospholipid antibodies were positive at 17U/ml. The Total hemolytic complement was normal at 32.8 with complement C3 decreased to 0.80g/l and complement C4 normal at 0.11g/l. The diagnosis of systemic lupus erythematosus with joint determinism was maintained. The evolution was favorable with corticosteroids and hydroxychloroquine.

Observation 2:

A 14-year-old schoolgirl with a history of recurrent angina pectoris, without any particular family history, received for diffuse pain evolving for 2 months in a context of fever, alteration of the general state (asthenia, weight loss, anorexia), photophobia and skin lesions located on the uncovered parts of her body. The clinical examination noted: a polyarthritis of the small and large joints with 52 painful joints and 5 swollen joints (wrists, right knee and ankles), and a cervical-thoracic-lumbar spinal syndrome. Scaly and crusted lesions in the shape of butterfly wings on the face and forearms(FiguresI and II); purpuric lesions on the palmar surfaces of the hands and ulcerations in the mouth. There were also multipleadenopathies in the cervical, submandibular, axillary and inguinal areas. The biological examination showed an anemia of 7.5 g/dl and a 24-hour proteinuria of 1.19 g/24h. The rest of the hematological, renal and hepatic investigations were normal. Immunologically, anti-nucleosome antibodies were positive at 215 U/L, anti-native DNA at 225 U/L, anti-histone at 220 U/L. Anti-ENA was positive with anti-SmD1 at 222 U/L, anti-RPP/Po at 220 U/L, anti-SSA/Ro at 125 U/L, anti-SSB/La at 75 U/L, anti-U1nRNP at 222 U/L. The diagnosis of systemic lupus erythematosus with joint, skin and renal involvement was maintained. The evolution was favorable under corticotherapy and hydroxychloroquine.

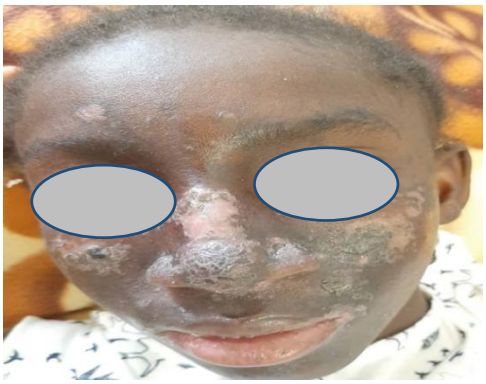

Figure I: Scaly and crusted lesions in the shape of butterfly wings on the face and forearms

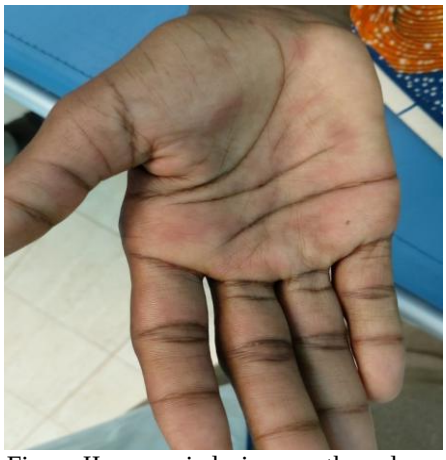

Figure II: purpuric lesions on the palmar

Observation 3:

11-year-old schoolgirl patient, first twin, HIV-positive type 1,in regular caresince 18 months of age with undetectable viral load on abacavir-dolutegravir-ritonavir. Mother and twin sister are HIV-positive and treated. She was followed up in pediatrics for bifocal tuberculosis (pulmonary and digestive), selected because of: prolonged fever, dypnea, polyseritis and generalised pruritus, all evolving in a context of altered general condition without other associated general signs. She was referred to the rheumatology department because of the persistence of the symptoms. Clinical examination revealed: alopecia, melanoderma, bluish nails, curly, red and brittle hair, suggestive of silky tricopathy and polyseritis. Investigations revealed anemia at 9.9 g/dl and 24-hour proteinuria at 0.885 g/24h. The rest of the haematological, renal and hepatic examination was normal. Immunology showed positive anti-nucleic acid antibodies at 1280 U/ml, positive anti-native DNA antibodies at 380 IU/ml and anti-ENA antibodies with anti-Sm(SmD3) at 33U/ml. The diagnosis of systemic lupus erythematosus with pulmonary and renal involvement in HIV was maintained. The evolution was favorable under corticotherapy and hydroxychloroquine for about 6 months. She died a few months later in a table of acute respiratory distress.

III. Discussion

Systemic lupus erythematosus (SLE) in children is a rare disease. The incidence of lupus varies between 0.36 and 0.60 per 100,000. The prevalence is estimated to be 5 to 10 per100,000 children. The disease usually begins in adolescence or sometimes in the prepubertal period. Onset of the disease before the age of two is very rare and often more severe with a higher frequency of cardiovascular, pulmonary and hematological involvement [3,4]. A retrospective study conducted in Nigeria over 4 years reported 12 cases of pediatric lupus. Of the 12 patients studied, eight were girls and four were boys. All patients had antinuclear antibodies and ENA antibodies. The anti-native ENA antibody was positive in 10 patients [5]. In French-speaking Africa, a case of nephrotic syndrome was reported in a 14-year-old girl in Mali, and autoantibodies, in particular anti-nucleic acid, anti-native DNA and anti-ENA antibodies, were detected. [6]. Our study is the first of its kind in French-speaking Africa to report, to our knowledge, 3 cases of pediatric lupus after 14 years of rheumatological practice in Burkina Faso [7]. The clinical picture is polymorphic with an unpredictable natural history [3]. Indeed, in the three observations we made, joint involvement was noted in two patients, as well as renal involvement. One patient had mucocutaneous involvement with erythema malarum and oral ulcerations associated with lymphadenopathy. Pulmonary involvement with polyseritis was seen in one patient. An association between lupus and HIV was noted in one of our patients; this association has been reported in some studies in adults [8,9]. Anti-Sm and anti-U1RNP antibodies are also frequently present [3]. In the immunological examination of our patients, anti-Sm was positive in two patients, as was anti-U1RNP. Treatment is a real challenge as there are no specific paediatric recommendations for management. All our patients were treated with a combination of corticosteroid therapy (1mg/Kg/D) and hydroxychloroquine (6.5mg/Kg/D). The severity of the disease and the need for daily treatment are unpredictable, making it very difficult for the child, and especially the adolescent, to comply with treatment. It is essential to involve the family and the child in a joint care system. It brings together paediatricians, psychologists, school doctors (in the form of an individualised reception project) and dieticians. The effect of corticosteroid therapy on body image, the impact of the disease on school life and social integration can be the cause of major depressive syndrome. Non-compliance is one of the elements responsible for relapse and is almost inevitable in adolescence [3]. The evolution of two of our patients has been favorable.

IV. Conclusion

The more severe pediatric form of SLE should be recognized and treated early. Morbidity and mortality depend on the organs involved. The severity of pediatric lupus requires early diagnosis and treatment to ensure the best control of the inflammation and to prevent the associated morbidity and mortality. Treatment relies on a multidisciplinary management to meet the challenges of a satisfactory statural growth of the child.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Abbreviations:

SLE: Systemic lupus erythematosus;
HIV: Human immunodeficiency virus;
ANA: Anti- nuclear antibody;
DNA: deoxyribonucleic acid

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Authors' contributions:
ATI collected and reviewed patient data and drafted the manuscript. YWN and OM conceived of the study and designed and coordinated the study and helped draft the manuscript. All authors critically revised the manuscript and read and approved the final manuscript.

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Declarations Ethics approval and consent to participate:
This case report was approved by the Institutional Review Board of Rheumatology Department of Bogodogo University Hospital.

Conflict of interest:
None

Consent for publication:
We have obtained the consent of the patients' guardians.

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