

Abstract citation ID: keac496.043

#### 47 SYSTEMIC LUPUS ERYTHEMATOSUS IN TUNISIAN CHILDREN: A CASE SERIES

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##### Background

Childhood-onset systemic lupus erythematosus (c-SLE) is the prototype of a multisystemic, inflammatory, heterogeneous autoimmune condition, characterized by simultaneous or sequential organ involvement. Compared with the adult onset form, c-SLE is thought to have a worse prognosis.

##### Objectives

To review the epidemiological, and bio-clinical, characteristics of a c-SLE case series.

##### Methods

The files of patients diagnosed as c-SLE in the pediatrics department of Monastir, Tunisia from January 2004 to March 2022 were reviewed. Mean and standard-deviations were used to express normally-distributed variables, as verified by the Kolmogorov-Smirnov statistical test.

##### Results

Fourteen patients were collected. Female to male ratio was 6:1. Mean ages at lupus onset and diagnosis were  $9.9 \pm 1.4$  years, [5–13.8 years] and  $10.75 \pm 2.3$  years [6–14 years], respectively. Only two children had a family history of autoimmune disease.

The initial admission was motivated primarily by skin and musculoskeletal manifestations, in 64.3% and 51.7% of cases, respectively. General signs (fever, asthenia) were observed in 35.7% of cases, hematological and gastrointestinal manifestations in 28.6% of cases each. In 3 cases, upper gastric endoscopy was performed prior to admission, in view of abdominal pain and vomiting.

The physical examination noted various anomalies. Malar rash (50%) and discoid lupus (28.6%) were the most frequent cutaneous manifestations, while skin biopsy was performed in three cases, all favorable for lupus. The musculoskeletal manifestations comprised arthralgia (71.4%), arthritis and myositis (14.3% each). Hematological manifestations included thrombocytopenia and leukopenia in 4 cases each, as well as 3 cases of auto-immune hemolytic anaemia and splenomegaly. Renal manifestations were dominated by proteinuria in 7 cases, followed by hematuria in 6 cases, and 2 cases of hypertension (associated with renal failure in one case). The renal biopsy was performed in one case showed a stage-2 lupus nephritis. Pleural effusion was observed in 3 cases, 3 cases of pneumonia, 2 cases of pericarditis, one case of myo-pericarditis and one case of central nervous system (CNS) lupus.

Relevant results of the laboratory workup are illustrated in the following table:

47 TABLE 1

Laboratory test	Performed (%)	Abnormal results (%)
Erythrocyte sedimentation rate	14 (100%)	13 (92%)
Antinuclear anti-bodies	14 (100%)	13 (92%)
AntiDNA antibodies	7 (50%)	5 (71%)
AntiSm antibodies	8 (57%)	5 (62%)
Antiphospholipid antibodies	8 (57%)	5 (62%)
Complement	11 (78%)	9 (81%)

The formal diagnosis of SLE was established according to the ACR-1997 criteria in 7 cases (50%), the SLICC-2012 in 4 cases (28.6%) and EULAR/ACR-2019 in 3 cases (21.4%). The c-SLE diagnosis was associated with coeliac disease and Hashimoto thyroiditis in one case each.

The therapeutic management was based on corticosteroids in 11 cases, followed by hydroxychloroquine in 3 cases, while cyclophosphamides and immunoglobulin were used for one case each.

The outcomes were heterogeneous. Among 11 patients with sufficient follow-up, 6 cases of remission and 2 cases of relapse were noted. Major adverse events were not infrequent: one case each of cardiac tamponade, macrophage activation syndrome and severe CNS lupus were observed, all fatal.

##### Conclusion

Childhood-onset systemic lupus is a challenging disease, both to diagnose and to treat. The development of new criteria of higher specificity and sensitivity has greatly helped identify the incomplete types of lupus and allow for early stage diagnosis, and therefore prevent the serious complications of the disease.