ANIFESTATIONS OF CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS: RESULTS OF SINGLE CENTER RETROSPECTIVE STUDY

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Background

Approximately 10-20% of patients (pts) with SLE develop the disease before 18 years old. Childhood-onset SLE (cSLE) usually has more aggressive course, perhaps due to the delay with diagnosis verification . The achievement of medication-free remission in adulthood is extremely rare in cSLE and quality of life remains compromised.

Objectives

To establish the specific features of cSLE at disease onset by the retrospective study in single center.

Methods

216 pts with cSLE who were hospitalized in our center from 1992 to 2017 were included in retrospective study.
Diagnosis of SLE was reviewed according to 2012 SLICC criteria. Clinical, hematological and immunological manifestations of SLE were evaluated. SLEDAI 2K was used for disease activity assessment.

Figure 1. Characteristics of the trigger`s factors (n=63)

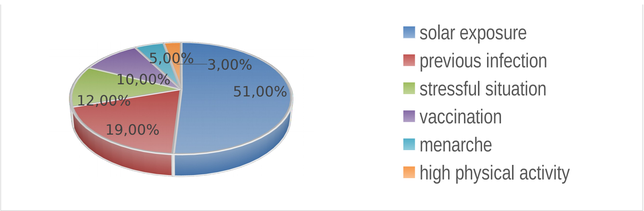


Table 1. Demographic characteristics of cSLE patients (n=216)

Characteristics	
Boys/girls	28/188
Age at the onset , Me (25;75)	13.7 y (10.8; 15.05)
Disease duration at the time of cSLE verification, Me (25:75)	6.0 mo (2.0;14.0)

Figure 2 Clinical features, %

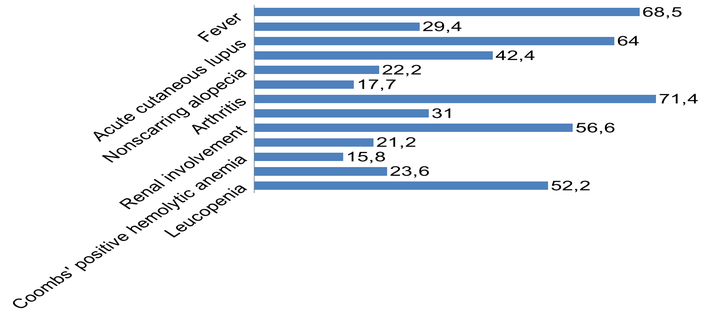
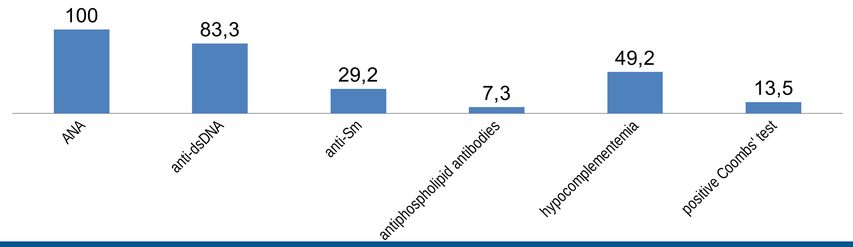


Figure 3. Immunological features, %



Results

12.9% of cSLE pts were boys (girls to boys ratio was 6.7:1). The triggering factor was found in 29% pts (Fig.1).Only 9.2% of patients initially had SLE diagnosis, 1.4% had discoid lupus erythematosus as an initial diagnosis, 21% - different infections, 11.2% - allergic diseases, 6.3% - nephritis, 33.6% - various rheumatic diseases (16.8% - juvenile idiopathic arthritis), in remaining 17.3% cases the information about initial diagnosis was missing. The median age at the onset was 13.7 y (10.8; 15.05); the median disease duration at the time of cSLE verification was 6 months (2; 14). In 33.4% pts cSLE was verified after 1 year disease duration, in 15.3% - after 3 years. The most common feature was arthritis – in 71.4%. Fever observed in 68.5% pts at the onset, significant weight loss – in 29.4%. 64% pts had acute cutaneous lupus at the onset, 42.4% - chronic cutaneous lupus, 17.7% - oral and nasal ulcers, 22.2% - nonscarring alopecia, 31% - serositis, 56.6% - renal involvement, 21.2% –neuropsychiatric disorder. The Coombs' positive hemolytic anemia was found in 15.8% pts, leucopenia/lymphopenia – in 52.2%, thrombocytopenia – in 23.6%. ANA were detected in 100% pts, anti-dsDNA – in 83.3%, anti-Sm – in 29.2%, antiphospholipid antibodies - in 7.3%, hypocomplementemia – in 49.0%, positive direct Coombs test out of hemolytic anemia – in 13,5 %. Macrophage activation syndrome at the onset was documented in 3.4% pts. Median disease activity by SLEDAI at the time of cSLE verification was 13.7 scores (8;20), maximum – 42.

Conclusion

cSLE presentation with non-specific general and constitutional manifestations in the majority of cases misled to erroneous interpretation of the condition as infectious or allergic disease in 1/3 of all cases. A monosymptomatic manifestation at the onset, such as arthritis, skin lesion or hematologic disorders, can lead to late diagnosis and very high activity at the moment of start therapy. Specific features of cSLE must be suspected in all cases of arthritis with skin lesions and/or any hematological manifestations, even non-specific.