Greater diagnostic delay in early-onset than in late-onset systemic lupus erythematosus - data from Reuma.pt/LES



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Introduction

Systemic lupus erythematosus (SLE) affects predominantly women of reproductive age. However, in about 15% of patients SLE begins before the age of 18 years (early-onset) and in 10-20% of patients SLE is first diagnosed after the age of 50 years (late-onset). The age at disease onset significantly impacts on clinical presentation, disease course, response to treatment and prognosis.

Objectives and Methods

- o To compare clinical and laboratory features of patients with earlyonset and those with late-onset SLE
- To determine whether age of onset is related to delay of diagnosis
- SLE patients (ACR criteria) from the registry Reuma.pt with disease onset at age≤18 years-old or at age≥50 years-old were included
- Cross-sectional analysis based on last recorded visit. Student's ttest, chi-square or Fisher's exact tests were carried out to compare the groups.

SLE patients in the register Reuma.pt/LES 1510 patients	SLE Classification (≥ 4 ACR criteria) 1296 patients	Age ≤ 18 years + ≥ 50 years 313 patients	Age ≤ 18 years (157 patients) Age ≥ 50 years (156 patients)
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Results

The Portuguese register of lupus patients - Reuma.pt/LES

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	Nº of participating centers	Nº of patients registered	Nº visits/patient:
December 2013	50	1510	2.06±2.28

Early-onset SLE:

- Diagnosis delay significantly larger (table 1);
- Longer disease duration (table 1);
- Anti-Sm positivity more frequent (table 1);
- Higher disease activity (table 1);
- Mycophenalate mophetil more commonly used (table 3).

Late-onset SLE:

- Photosensitivity, arthritis and neurological disorder more prevalent (table 1);
- Greater accumulated damage (table 1);
- Comorbidities more common (table 2).

Characteristics	Early onset SLE	Late onset SLE	P
Characteristics	(n= 157)	(n=156)	
Men	19 (12.1 %)	29 (18.6 %)	0.111
Caucasians	140 (94.6%)	121 (97.6)	0.418
Age at disease onset (years)	13.9±3.0	58.9±7.2	NA
Diagnosis delay (years)	3.1±5.03	1.68±3.05	0.001
Disease duration (years)	16.8±10.5	9.37±5.2	<0.001
Education (years), n=54	10.3±3	6.5±4.33	<0.001
Malar rash, n=238	56 (44.4%)	51 (45.5%)	0.866
Discoid rash, n=240	6 (4.7%)	9 (8.0%)	0.221
Photosensitivity, n=241	50 (39.1%)	62(54.9%)	0.014
Oral ulcers, n=239	33 (26.0%)	38 (33.9%)	0.180
Arthritis, n=241	86 (67.2%)	92 (81.4%)	0.012
Serositis, n=241	36 (28.1%)	26 (23.0%)	0.365
Renal Involvement, n=237	34 (27.0%)	26 (23.4%)	0.529
Neurologic disorder, n=239	2 (1.6%)	9 (8.1%)	0.017
Hematologic disorder, n=240	78 (60.9%)	71 (63.4%)	0.696
Anti-DNA, n=237	93(73.2%)	81 (73.6%)	0.943
Anti-Sm, n=232	21 (16.4%)	6 (5.8%)	0.009
Anti-cardiolipin, n=224	32 (26.6%)	21 (21.0%)	0.400
SLEDAI-2K*, n= 252	3.0±3.3	2.0±2.8	0.01
SLICC damage index, n=252	0.69±1.4	1.0±1.33	<0.001

Table 1 – Demographic and clinical characteristics of SLE patients

^{*} At the last recorded visit; n = number of patients evaluated

Comorbidities	Early onset SLE (n= 157)	Late onset SLE (n=156)	P
Hypertension, n=149	15 (24.6%)	41 (46.6%)	0.006
Diabetes, n=252	1 (0.8%)	9 (6.9%)	0.013
Thyroid disease, n=149	2 (3.2%)	17 (19.3%)	0.003
Anti-phospholipid syndrome,	8 (13.1%)	3 (3.4%)	0.026
n= 149			
Sjogren's syndrome, n=149	3 (11.0%)	13 (14.8%)	0.047

Table 2 – Comorbidities of SLE patients n = number of patients evaluated

Medication	Early onset SLE (n= 157)	Late onset SLE (n=156)	P
Antimalarial drugs (ever)	109 (69.4%)	113 (72.4%)	0.589
Corticosteroids (ever)	95 (60.5%)	91 (58.3%)	0.366
Other Imunossupressants (ever)			
- Azathioprine	42 (26.8%)	32 (20.5%)	0.111
- Methotrexate	10 (7.0%)	19 (12.2%)	0.924
- Cyclophosphamide	5 (3.2%)	2 (1.3%)	0.082
- Cyclosporine	4 (2.5%)	2 (1.3%)	0.135
- Mycophenalate mophetil	35 (22.9%)	5 (3.8%)	<0.001
Biological therapies (ever)	8 (5.1%)	4 (2.6%)	0.092

Table 3 – Medication used for SLE patients

n = number of patients evaluated

Conclusions

- Age of onset is associated with a significant difference in clinical characteristics, disease outcome and time until diagnosis.
- Diagnosis delay is significantly higher in early onset disease, which could be explained by oligosymptomatic or atypical SLE manifestations.
- Diagnosis delay may negatively impact the prompt control of SLE, essential to ensure normal development in childhood and adolescence.