Demographic data of Egyptian juvenile systemic lupus erythematosus during COVID-19 pandemic: A single centre study

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Abstract---Background: Juvenile Systemic lupus erythematosus is a chronic autoimmune inflammatory disease that causes damage to various organs such as kidneys, blood, musculoskeletal and nervous system. SLE is variable ranging from mild to severe with unpredictable course of the disease and periods of remission and flares. Methods: The study included 67 patients diagnosed as having jSLE according to the Systemic Lupus International Collaborating Clinics (SLICC) for whom demographic data, full history and laboratory investigations were collected. Results: Among our study population were 13.4% males and 86.6% females with male to female ratio (1:6.4). Their mean age was 10.94±3 years. Most of our patients were from Giza (43.3%) followed by Cairo (22.4%). C3 and C4 were consumed in 74.6% of patients, proteinuria was present in 73.1% of patients, hematuria was present in 25.4% of patients. ANA was positive in 98.5% of patients at the time of diagnosis while antiDNA was positive in 85.1%. Conclusion: Juvenile systemic lupus
erythematous is not uncommon in Egypt with female predominance as known worldwide but younger age of onset of the disease. During COVID 19 pandemic follow up was limited due to fear of infection.

**Keywords**—juvenile systemic lupus erythematosus, antiphospholipid syndrome, proteinuria.

**Introduction**

Juvenile Systemic lupus erythematous (JSLE) is a chronic autoimmune inflammatory disease that causes damage to various organs such as kidneys, skin, blood cells, musculoskeletal and nervous system. SLE is variable ranging from mild to severe with unpredictable course of the disease and periods of remission and flares (Mohan and Puttermann, 2015). Overall prevalence of SLE in the general population vary worldwide ranging from 4 to 45/100000 in Asia-Pacific countries, to 52–150/100000 in the USA being greater in non-white races (Gaballah et al., 2019). The prevalence is influenced by ethnicity, where African-Americans /African Caribbeans and Hispanics are three times as likely to have the disease compared to European-Caucasians (Mousa et al., 2018).

Juvenile systemic lupus erythematosus (jSLE) manifests before the 18th birthday. Peak age of onset is approximately 12.6 years (Smith et al., 2019). Patients with disease onset before 5 years of age are uncommon and referred to as early-onset SLE (Hedrich et al., 2017). In pediatrics, female to male gender distribution ranges around 5:1. It is even close to 1:1 before 5th year of age, while the predominance in females rises to 9–10:1 in adult SLE, this may be related to the influence of hormonal factors on disease pathogenesis (Smith et al., 2019) (Watson et al., 2012). The pathophysiology of SLE is not fully understood.

**Patients and methodology**

Our study was conducted on patients following up at Pediatric Rheumatology Unit, Cairo University Pediatric Hospitals, Cairo, Egypt during the period of COVID 19 pandemic. The study included 67 patients diagnosed as having jSLE according to the Systemic Lupus International Collaborating Clinics (SLICC) (Petri et al., 2012).

**For the 67 patients the baseline data were collected as follows**

1. Demographic date:
   - age
   - sex
   - residence
2. Full history including
   - age at disease diagnosis
   - age at onset of disease presentation
   - positive family history of rheumatological disease.
3. Laboratory investigations at time of study conduct and diagnosis including;

- CBC with differential
- ESR
- serum C₃, C₄
- anti-dsDNA
- creatinine,
- urine analysis
- albumin/creatinine ratio

**Results**

Our study included 9 males (13.4%) and 58 females (86.6%) with JSLE. Their mean age was 10.94±3 years. Mean age at first presentation of the disease was 8.69±3 years, mean age at diagnosis was 8.87±2.9 years with mean disease duration of 2.25 years with minimum duration of 6 month of follow up. Most of our patients were from Giza 43.3% followed by Cairo 22.4% then 10.4% from Fayoum. The remaining 23.9 % were distributed over other governemets as Elminia, Aswan, Banisweef, Hurghada, Sharkia, Tanta, Upper Egypt, Kalyobia and Monofia.

![Figure (1) pie chart showing geographic distribution of JSLE patients in Egypt](image)

Nine patients have family history of SLE (13.4%) while 3 patients have family history of other rheumatological disease as juvenile idiopathic arthritis and familial Mediterranean fever (4.5%).
Laboratory findings at the time of disease presentation

Initial laboratory investigations at the initial disease presentation showed mean hemoglobin of 9.4 ±2.52 g/dl, total leucocytic count of 8.15±4.57 ×10^3/cmm, platelet count 230.4±127.7 ×10^3/cmm, ESR 42.5 ±24.6mm/hour, creatinine 0.66±0.29 mg/dl and Albumin/creatinie ratio in urine 1002.57±1504.86 mg/g creat.

Table (1) laboratory findings at time of disease presentation

<table>
<thead>
<tr>
<th>Cases</th>
<th>Mean</th>
<th>SD</th>
<th>Median</th>
<th>Minimum</th>
<th>Maximum</th>
</tr>
</thead>
<tbody>
<tr>
<td>HB</td>
<td>9.40</td>
<td>2.52</td>
<td>9.00</td>
<td>4.50</td>
<td>15.00</td>
</tr>
<tr>
<td>TLC</td>
<td>8.15</td>
<td>4.57</td>
<td>7.00</td>
<td>2.00</td>
<td>23.00</td>
</tr>
<tr>
<td>PLT</td>
<td>230.40</td>
<td>127.78</td>
<td>234.00</td>
<td>0.00</td>
<td>528.00</td>
</tr>
<tr>
<td>ESR</td>
<td>42.52</td>
<td>24.66</td>
<td>40.00</td>
<td>10.00</td>
<td>130.00</td>
</tr>
<tr>
<td>C3/C4</td>
<td>0.66</td>
<td>0.29</td>
<td>0.60</td>
<td>0.20</td>
<td>1.80</td>
</tr>
<tr>
<td>A/C Ratio</td>
<td>1002.57</td>
<td>1504.86</td>
<td>495.00</td>
<td>14.00</td>
<td>8761.00</td>
</tr>
</tbody>
</table>

C3 and C4 were consumed in 50 patients (74.6%), proteinuria was present in 49 patients (73.1%), hematuria was present in 17 patients (25.4%). Anticardiolipin IgG was positive in 13 patients (19.4%) initially and IgM positive in 19.4% as well while lupus anticoagulant was positive 13 patients (19.4%). When repeated at least 12 weeks after, 6 patients (9%) were still positive and were diagnosed as antiphospholipid syndrome with no thrombotic events.

Table (2): laboratory findings at time of disease diagnosis

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hematuria</td>
<td>Present</td>
<td>17</td>
</tr>
<tr>
<td>Proteinuria</td>
<td>Present</td>
<td>49</td>
</tr>
<tr>
<td>C3</td>
<td>Consumed</td>
<td>50</td>
</tr>
<tr>
<td>Test</td>
<td>Result</td>
<td>Positive</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>-----------</td>
<td>----------</td>
</tr>
<tr>
<td>C4</td>
<td>Consumed</td>
<td>50</td>
</tr>
<tr>
<td>anticardiolipin IgG</td>
<td>Positive</td>
<td>13</td>
</tr>
<tr>
<td>Anticardiolipin IgM</td>
<td>Positive</td>
<td>13</td>
</tr>
<tr>
<td>lupus anticoagulant</td>
<td>Positive</td>
<td>13</td>
</tr>
<tr>
<td>ANA</td>
<td>Positive</td>
<td>66</td>
</tr>
<tr>
<td>antiDNA</td>
<td>Positive</td>
<td>57</td>
</tr>
</tbody>
</table>

Figure (3): bar chart showing percentage of antiphospholipid syndrome patients among the study population

ANA was positive in 66 patients (98.5%) of patients at the time of diagnosis and negative in only one patient (1.5%) while antiDNA was positive in 57 patients (85.1%).

Figure (4): bar chart showing ANA and antiDNA positivity among the study population
Discussion

Among our study population (67 patients) were 9 males (13.4%) and 58 females (86.6%) with male to female ratio (1:6.4), this comes in concordance with the results of the study of Torrente-Segarra et al., 2019 on Caucasian juvenile SLE patients, Sit and Chan 2018 on Chinese, Abdel-Nabi et al., 2018 on Egyptian SLE patients where male to female ratio ranged from 1:1.54 to 1:7.85 showing more prevalence in females. In our study, Mean age of the JSLE group was 10.94±3 years with mean age at time of diagnosis of the disease 8.69±3years and mean disease duration of 2.25 years. This was generally a younger age group with subsequently a shorter disease duration and follow up than most of the other studies, as in ElGarf et al., 2021, salah et al 2011 and Salah et al 2009 with mean age at diagnosis of 11.12 years and mean disease duration of 5.72 years.

In our cohort, 17.9% patients had a family history of one or more autoimmune disease, where 13.4% had positive family history of SLE and 4.5% had family history of other rheumatological disease. This comes in agreement with most of the studies done on younger patients with juvenile onset autoimmune diseases as in Ashournia et al., 2018. This could be explained by the fact that autoimmune diseases specially SLE has genetic background. Regarding the laboratory findings; Results of both ANA and Anti-DNA was very close in almost all studies where in our cohort ANA was positive in 98.5% of patients at the time of diagnosis and negative in only one patient (1.5%) while Anti-DNA was positive in 85.1%. Our results regarding autoantibody results were in concordance with many other studies like those of Mohamed et al 2018, mosaad et al 2015, Szymanik-Grzelak et al., 2021 as well as in adult studies as in Martens et al., 2009. Anti-DNA positivity was lowest in salah et al 2011 still ANA positivity was in 94.1%.

This ensures the importance of ANA positivity in diagnosis of SLE and justify its use as an entry criterion in 2019 European League Against Rheumatism (EULAR)/ACR SLE classification criteria (Aringer et al. 2019) In our study; Anti-phospholipid syndrome was diagnosed in 9% of the patients compared to 11.5% in Hadidi et al 2018, the reason for the difference is Hadidi et al study involves adults with other comorbidities. Laboratory findings of lupus nephritis were studied in our work, with complement consumption found in 74.6% of patients which was in concordance with the results of El-Garf et al., 2021 and Mohamed et al 2018. Proteinuria were detected in 73.1% of our study population similar to in Mohamed et al 2018, hematuria was detected in 31.2% of patients at time of disease diagnosis which was close to the work of Mohamed et al 2018 and El-Garf et al., 2021 but lower than Szymanik-Grzelak et al., 2021 (89.7%) as his study was conducted only on lupus nephritis patients.

References


