MJMR, Vol. 33, No. 2, 2022, pages (171-174).

Open Access
ISSN:2682-4558

Research Article

Hematological Parameters in Pediatric-onset Systemic lupus Erythematosus and its association with disease activity

Sheren Esam Maher [1], Suzan Mohamed Aly Omar [1], Mostafa Ahmed El Sayed [2], and Ahmed Ragab Abdul Badia[1]

[1] Department of Pediatrics, Faculty of Medicine, Minia University, El-Minia, Egypt.
[2] Department of Clinical Pathology, Faculty of Medicine, Minia University, El-Minia, Egypt.

Abstract

Background: Pediatric onset Systemic lupus erythematosus (P-SLE) is an autoimmune multisystem illness with disease occurrence before the age of eighteen. It is a highly complicated condition with a significant variability across individuals**.** Aim of the study: To assess hematological parameters in P-SLE patients attending to the Pediatric Rheumatology out-patient clinic in Minia University Children Hospital, as well as, admitted to the in-patient ward and to evaluate the relationship between these hematological parameters and activity of P-SLE disease. **Methods:** This study was a prospective cross-sectional study included 40 children diagnosed with SLE attending to the Pediatric Rheumatology out-patient clinic in Minia University Children Hospital, as well as, admitted to the in-patient ward, from August 2020 to January 2022. Also, 40 apparently healthy children were included as a control group. They are age and sex matched, with the children having SLE. All enrolled Patients were subjected to: Detailed history taking, complete clinical examination, laboratory investigations including: complete blood count (CBC) **Results**: Both Patients with mild disease activity and Patients with moderate to high disease activity had significantly lower hemoglobin level than controls (P= 0.004 and 0.04 respectively), but there is non significant positive correlation between Hb level and SLEAI score as r = 0.382 and p = 0.142. **Conclusion:** Children having Systemic lupus erythematosus are more likely to have lower hemoglobin level than healthy children which may result in high morbidity in P-SLE, but lower hemoglobin level may not be closely associated with P-SLE disease activity.

Keywords; Systemic lupus erythematosus; autoimmunity; Rheumatology; hematological disorders.

Introduction

Pediatric onset Systemic lupus erythematosus (p-SLE) is an autoimmune multisystem illness with disease occurrence before the age of eighteen.(1) It is a highly complicated condition with a significant variability across individuals.(2)

P-SLE is usually a more serious disorder involving renal, neurological and hematological systems, resulting in lifelong damage compared to adult-onset lupus patients(3).
Pediatric-onset SLE has a widely diverse presentation and clinical outcomes.

Consequently, diagnosis might be problematic. It ranges from relatively mild illness to severe, life-threatening manifestations. Countless symptoms, such as headaches, oral ulcers, arthralgia, fever and weight loss, are non-precise and occur in children for a diversity of reasons. Major organ involvement is dominant, with renal affection being the most common. Lupus nephritis (LN) is present in up to 80% of patients, with up to 19% having end-stage renal failure(4-6).

Rheumatologists as case managers, dermatologists, nephrologists, hematologists, neurologists, radiologists, immunologists, gastroenterologists, cardiologists, endocrinologists, and infectious disease experts are typically required due to the complexity of p-SLE with multi-organ involvement (7).

Patients and methods

This study was a prospective cross-sectional study included 40 children diagnosed with Systemic Lupus Erythematosus attending to the Pediatric Rheumatology out-patient clinic in Minia University Children Hospital, as well as, admitted to the in-patient ward, from August 2020 to January 2022.

Also, 40 apparently healthy children were included as a control group. They are age and sex matched, with the children having SLE.

The Systemic Lupus Erythematous Disease Activity Index (SLEDAI) was calculated for all children with SLE. Activity categories have been defined based on SLEDAI scores

The subjects of the study were grouped accordingly into the following three groups:

- **Group (A)** [Mild activity]
  this group includes children with SLE with SLEDAI score less than or equal to 5.
- **Group (B)** [Moderate-high activity]
  this group includes children with SLE with SLEDAI score greater than or equal to 6.
- **Group (C)** [control]
  this group includes apparently heathy children**.**

Then included children were subjected to the following:

A- Careful history taking including:

Name, age, sex, residence, socioeconomic standard and family history of blood diseases.

B- ***Full clinical examination:*** including

1. Vital data: respiratory rate, heart rate, blood pressure, temperature.
2. 2- Systematic examination including
3. Chest
4. Heart
5. Abdomen
6. Musculoskeletal System and Joints
7. Neuropsychiatric assessment

C-***Laboratory investigations:***

1. Complete blood count (CBC)
2. Renal Function tests
3. Liver function tests
4. Urine analysis
5. C-Reactive Protein (CRP)
6. Erythrocyte Sedimentation Rate (ESR)

Results

Table 1: showed that group A had significantly lower hemoglobin level than controls (P =0.004).

Table 1: comparison between Group A (mild SLE disease activity), Group B (moderate to high disease SLE activity) & controls as regarding hematological data:

Baseline data	Group A n = 20	Group B n = 20	control n = 40	P value	
	Mean ± SD	Mean ± SD	Mean ± SD		
Age (years)	12.76 ± 2.46	11.88 ± 2.54	12.32 ± 2.51	0.502	
Hb (gm/dL)				0.008*	
				A Vs B	0.432
				A Vs C	0.004*
	10.25 ± 0.83	10.5 ± 1.03	11.0 ± 0.98	B Vs C	0.04*
TLC (x10 ^{3}cells /mm ^{3})	7.28 ± 1.98	7.09 ± 2.65	6.68 ± 1.42	0.166	
PLT (x10 ^{3} platelets /*mcL)*	260.2 ± 65	207. ± 96.2	229. ± 67.5	0.101	

Moreover, Group B had significantly lower hemoglobin level than control (p=0.04)

n = number, SD =standard deviation, Hb = Hemoglobin, TLC = Total Leucocyte Count, PLT= platelets

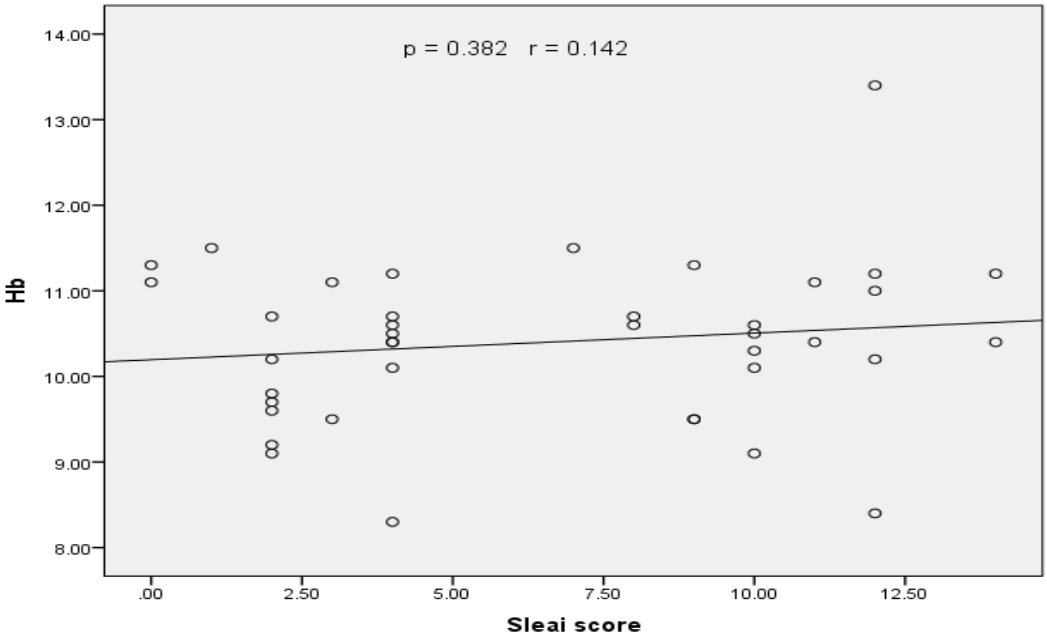

Figure (1): correlation between Hb level and SLEAI score.

Figure 1 showed non significant positive correlation between Hb level and SLEAI score as r = 0.382 and p = 0.142.

Discussion

Estimated incidences of p-SLE array among 0.36 and 2.5 per 100,000 children, with a prevalence of 1.89–34.1 per 100,000 children[8-10].
Hematological manifestations in systemic lupus erythematosus (SLE) are common and diverse. Their frequency varies in different populations.[11]
The major findings of our study is that; Patients with mild disease activity (group A) had significantly lower hemoglobin level than controls (P= 0.004). Moreover, Patients with moderate to high disease activity (group B) had significantly lower hemoglobin level than controls (P=0.04), but there is non significant positive correlation between Hb level and SLEAI score as r = 0.382 and p = 0.142.

This was in agreement with Samohvalov et al., (2018) who found that anemia is present in approximately half of the people with active Systemic lupus Erythematosus.

Moreover, our results were in agreement with Giannouli et al., (2006) who found that hematological abnormalities are very common in systemic lupus erythematosus and anemia is found in approximately 50% of SLE patients.

On the other hand, our results were in disagreement with Samohvalov et al., (2012) who found that Anemia is closely associated with disease activity.

Also, our results were in disagreement with Beyan et al., (2007) who found that patients with high disease activity had significantly lower hemoglobin level than patients with low disease activity (P=0.018).

These hematological manifestations can be due to the disease itself, another concomitant disease or iatrogenic. Hemolytic anemia, leukopenia, lymphopenia and thrombocytopenia are incorporated into both the 1997 update of the 1982 American College of Rheumatology (ACR). Anemia is frequent, affecting more than 50% of patients throughout the course of the disease and it can be both immune and nonimmune mediated in SLE patients. [12]
Anemia of chronic disease (ACD) is the most common type of anemia in SLE patients, responsible for about one third of the cases [13].

Autoimmune Hemolytic Anemia, Iron Deficiency Anemia, drug-induced myelotoxicity, and anemia due to chronic renal failure are also often detected.

Aplastic anemia, Pure Red Cell Aplasia, pernicious anemia, myelofibrosis, sideroblastic anemia, hemophagocytic syndrome occur less frequently [12-14]

Anemia is associated with high morbidity in different clinical conditions.
For example: developmental delay in children, changes in immunological status, high risk of infections. Anemia is an independent risk factor for the development of cardiovascular complications in general population [15].

Conclusion

Children having Systemic lupus erythematosus are more likely to have lower hemoglobin level than healthy children which may result in high morbidity in P-SLE, but lower hemoglobin level may not be closely associated with P-SLE disease activity.

References

1. Mosca, M., J.T. Merrill, and S. Bombardieri, Chapter 2 - Assessment of Disease Activity in Systemic Lupus Erythematosus, in Systemic Lupus Erythematosus, G.C. Tsokos, C. Gordon, and J.S. Smolen, Editors. 2007, Mosby: Philadelphia. p. 19-23.
2. Hedrich, C.M., E.M.D. Smith, and M.W. Beresford, Juvenile-onset systemic lupus erythematosus (jSLE) - Pathophysiological concepts and treatment options. Best Pract Res Clin Rheumatol,2017.**31**(4):p.488-504.
3. Tan, J.H., et al., Childhood-onset systemic lupus erythematosus in Singapore: clinical phenotypes, disease activity, damage, and autoantibody profiles. Lupus, 2015. **24**(9): p. 998-1005.
4. Al-Mayouf, S.M., et al., Outcome of childhood lupus nephritis in Saudi children. Saudi J Kidney Dis Transpl, 2017. **28**(5): p. 1015-1020.
5. Samanta, M., et al., Childhood lupus nephritis: 12 years of experience from a developing country's perspective. Eur J Rheumatol, 2017. **4**(3): p. 178-183.
6. Jebali, H., et al., Clinicopathological findings and outcome of lupus nephritis in Tunisian children: a review of 43 patients. Pan Afr Med J, 2017. **27**: p. 153.
7. Groot, N., et al., European evidence-based recommendations for diagnosis and treatment of childhood-onset systemic lupus erythematosus: the SHARE initiative. Ann Rheum Dis, 2017. **76**(11): p. 1788-1796.
8. Pineles, D., et al., Worldwide incidence and prevalence of pediatric onset systemic lupus erythematosus. Lupus, 2011. **20**(11): p. 1187-92.
9. Hiraki, L.T., et al., Prevalence, incidence, and demographics of systemic lupus erythematosus and lupus nephritis from 2000 to 2004 among children in the US Medicaid beneficiary population. Arthritis Rheum, 2012. **64**(8): p. 2669-76.
10. Rees, F., et al., The incidence and prevalence of systemic lupus erythematosus in the UK, 1999-2012. Ann Rheum Dis, 2016. **75**(1): p. 136-41.
11. Alarcón, G.S., et al., Systemic lupus erythematosus in three ethnic groups. XIX. Natural history of the accrual of the American College of Rheumatology criteria prior to the occurrence of criteria diagnosis. Arthritis Rheum, 2004. **51**(4): p. 609-15.
12. Velo-García, A., S.G. Castro, and D.A. Isenberg, The diagnosis and management of the haematologic manifestations of lupus. J Autoimmun, 2016. **74**: p. 139-160.
13. Voulgarelis, M., et al., Anaemia in systemic lupus erythematosus: aetiological profile and the role of erythropoietin. Ann Rheum Dis, 2000. **59**(3): p. 217-22.
14. Giannouli, S., et al., Anaemia in systemic lupus erythematosus: from pathophysiology to clinical assessment. Ann Rheum Dis, 2006. **65**(2): p. 144-8.
15. Samohvalov, E. and S. Samohvalov, The Pattern of Anemia in Lupus. 2018.