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Original Article

Heamatologic manifestation of pediatric systemic lupus erythematosus at Queen Rania hospital for children

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ABSTRACT

Aim: The aim of this retrospective study was to highlight the spectrum of heamatological manifestations in PSLE at Queen Rania Hospital for children(QRHC). Methods: This is a retrospective study that included 25 pediatric patients ,diagnosed with PSLE over a period of 11 years from January 2000 till December 2010 at QRHC at King Hussein Medical center ,Amman Jordan. Medical records were reviewed for heamatological findings including complete blood count,coombs test ,antiphospholipid antibodies ,anticardiolipin antibodies,Anti-neuclear anti bodies 7and Anti ds-DNA. Results: Heamatological abnormities ,that include abnormalities in complete blood count were detected in 48%at the time of diagnosis,which included leucopenia ,hemolytic anemia in 24%and 20% respectively and thrombocytopenia and lymphopenia in 16% for each. Positive ANA,and anti ds DNA were found in 100% and 88% respectively.Normal platelets were found in 74% while high platelets just in 10%.High erythrocyte sedimentation rate (ESR) was found in 84%(21). Anti cardiolipin and lupus anticoagulant in 40%(10)and 16%(4) respectively. Conclusion: heamatologic manifestation in pediatric SLE was found In almost 50% of cases ,but when high ESR is added that will increase the percentage to more than 80%of cases. hematologic finding including leucopenia,anemia,lymphopenia,and high ESR could correlate with the activity of the SLE.

INTRODUCTION

Systemic lupus erythematosus(SLE) is a chronic multisystem autoimmune disease characterized by multiorgan involvement ,and presence of numerous auto-antibodies.

Pediatric SLE(PSLE) represents 10-20% of all SLE cases which associated with high disease activity[1].

The disease has variable presentation including constitutional symptoms ,serositis ,arthritis cutaneous ,pulmonary renal ,neurologic and heamatologic manifestations [2].

Heamatological findings are common in pediatric SLE ,it has been reported in as high as in 86% of the PSLE patients[3].

The aim of this retrospective study was to highlight the spectrum of heamatological manifestations in PSLE at Queen Rania Hospital for children(QRHC).

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METHODS

January 2000till December 2010 at QRHC at King Hussein Medical center ,Amman Jordan.

All patient should be below the age of 14 years, and should meet four or more of the revised American College of Rheumatology (ACR)classification criteria[4].

All patients charts were analyzed for demographic ,clinical and laboratory findings .

Medical records were reviewed for heamatological findings , including complete blood count,coombs test, antiphospholipid antibodies ,anticardiolipin antibodies,Anti-neuclear anti bodies and Anti ds-DNA.

Revision was done from the onset of the disease till the end of the study date.

Heamatological involvement was defined as 1.Hemolytic anemia2. Thrombocytopenia, less than 150X109/l,Leukopenia <4X109/l,Lymphopenia ,less than age related lower limit of normal , ANA more or equal to 1:80.

.This is a retrospective study that included 25 pediatric patients ,diagnosed with PSLE over a period of 11 years from

RESULTS

A total of 25 patient were included The mean age at onset of diagnosis was 10.9 years ,with PSLE developing in 80% after the age of 10 years .The female to male ratio was 7.3:1.The mean time from the start of illness to diagnosis was 8.6 month(range1-36)as shown in Table 1.

Non classical (non diagnostic) features were seen in almost all patients ,fatigue in 84%(21),anorexia 72%(18)and fever in 28% (7)of patients.at presentation cutaneous ,and arthritis were found in 68%(17) and 60%(15) respectively, while renal and neurological were found in 40%and 24 respectively ,as shown in Table 2.

Heamatological abnormities ,that include abnormalities in complete blood count were detected in 48%at the time of diagnosis,which included leucopenia ,hemolytic anemia in 24%and 20% respectively and thrombocytopenia and lymphopenia in 16% for each.

Normal platelets were found in 74% while high platelets just in 10%.

High erythrocyte sedimentation rate (ESR) was found in 84%(21).

Positive ANA,and anti ds DNA were found in 100% and 88% respectively.ANA was equal or more than 1:640 in more than 80% of cases.

Anti cardilipin and lupus anticoagulant in 40%(10)and 16%(4) respectively.

Pancytopenia in 3 patient who underwent bone marrow aspiration ,which revealed reactive ,and non showed findings suggestive macrophage activation syndrome.

Table -3 showed the heamatological abnormalities.

Table -1 Patient's data

Data		Number(%)
Number of patients		**25**
Gender	**female**	**22(88%)**
	male	**3(12%)**
	Ratio	**7.3:1**
Mean age at onset		**10.9 years**
Mean time of diagnostic delay		**8.6 months**

Tabl-2 clinical data

Clinical findings	Number and% of patients
Fatigue	21(84%)
Anorexia	18(72%)
Cutaneous	17(68%)
Arthritis	15(60%)
Renal	10(40%)
Fever	7(28%)
Neurologic	6(245)

Table -3 Heametologial manifestations

Hematologic data	Number and %
Anemia	8(32%)
Coombs positive Hemolytic anemia	5(20%)
Leucopenia	6(24%)
Leukocytosis	3(12%)
Lymphopenia	4(16%)
Thrombocytopenia	4(16%)
Pancytopenia	3(12%)
High ESR	21(84%)
Positive ANA	25(100%)
Anti ds DNA	22(88%)
Anticardiolipin antibodies	10(40%)
Lupus anticoagulant	4(16%)
CRP	4(16%)

Discussion

In our cohort demographic data ,age at presentation , female to male ratio and clinical presentation is comparable to the PSLE behavior in our region and other parts of the world[5].

Heamatologic manifestations are common in pediatric SLE and often a presenting feature of PSLE, anemia being the most common heamatologic manifestation followed by leucopenia [6,7].

In this retrospective analysis of 25 patients with a diagnosis of PSLE at initial presentation heamatologic involvement found in almost 50% of cases.Anemia was the most common presenting heamatologic finding in our patients 32% ,which was heamolytic in 20%, and non hemolytic in 12% including nutritional,anemia of chronic disease and others,which is consetent with other reports [3,8].

Direct coomb's test positive hemolytic anemia was associated with sever presentation ,as it was noted by other authors [9,10].

Leucopenia is common in PSLE due to lymphopenia ,neutropenia or both [11].

Leucopenia was the second most common heamatologic feature of PSLE at initial presentation in our cohort 24% ,while lymphopenia was in 16% . leucopenia and lymphoenia has been noted in 20% of patients ,especially in those with active disease [9].

Leukocytosis is as usual not found in uncomplicated SLE but mostly might indicate the associated underlying infectious process. Leukocytosis was detected in 2 patients with intercurrent infection and one patient who was on steroid before establishment of the diagnosis.

Thrombocytopenia is a common and important presentation of SLE, which has a direct relation with its morbidity and mortality [12].

In our study 16% showed thrombocytopenia while 74% showed normal platelets count .

Four patient were given a diagnosis of idiopathic thrombocytopenic purpura,one of them underwent splenoectomy 3 years before the diagnosis of SLE was confirmed ,the same finding was noted by other study [13].

High ESR was found in 84% of our cohort with a median of 65mm/hour, and the level was correlated with the activity of the disease in contrast with the CRP which was positive in just four patient (16%),which is expected as it is not correlate with the disease activity, this finding was compactable to what been found in a French multicenter study [14].

ANA and Anti ds DNA were positive in 100%,and 88% respectively with a mean of ANA of 1:640,this is consistent with Saudi report [8]. but different from what been reported by Meuner et al [15] , while positive anti ds DNA is almost as the same as the report by Tadio et al[16] Non of our patient had a negative ANA at presentation .

The available at our laboratory two antiphospholipid antibodies were done for all patients and they were positive in 40% for anticardiolpin antibodies and in 165for lupus anticoagulant.

presence of these antibodies found not only to increase the risk of development of thrombosis [17],but valvular abnormalities [18],and avascular bone necrosis [19].

Conclusion

heamatologic manifestations in pediatric SLE was found In 50% of cases ,but when high ESR is added that will increase the percentage to more than 80%of cases' .

Anemia was the commonest heamatologic finding .abnormal hematologic finding including leucopenia,anemia, lymphopenia,and high ESR could correlate with the activity of the SLE.

References

[1] kamphius S,Silverman ED.Prevelence and burden of pediatric-onset systemic lupus erythematosus.Nat Rev Rheumatol 2010;6:538-546.

[2] Brunner HI,Silverman ED,To T,Bombadiev c,Feldman BM.Risk factors for damage in childhood-onst systemic lupus erythematosus.Arthritis Rheum 2002;46:436-44.

[3] Gokce M,Bilginer Y,besbas N,Ozaltin F,cetin M,Gumruk F,et al.heamatologic features of pediatric systemic lupus erythematosus :suggesting management strategies in children.Lupus 2012;21:878-884.

[4] Hochberg MC.Updating the American college of Rheumatology revised criteria for classification of systemic lupus erythematosus .Arthritis Rheum 1997;40:1725.

[5] Abu-shukair M,Habahbeh Z,Almutereen m,Zyoud R,et al .spectrum of pediatric systemic lupus erythematosus at Queen Rania Al-Abdallah hospital for children.Accepted for puplication at medical journal of Jordanian Royal medical services.

[6] Pluchinotta FR,Schiavo,Vittadello F,et al. distinctive clinical features of pediatric systemic lupus erythematosus in three different age cases.lupus 2007;16(8):550-5.

[7] Hoffman IE,Lauwerys BR,De Keyer F,et al.Juvenile-onset systemic lupus erythematosus :different clinical and serological pattern than adult-onset systemic lupus erythematosus. Ann Rheum Dis 2009;68(30;412-5.

[8] Muzaffer MA,Al-Mayouf SM. clinical and laboratory variables of childhood systemic lupus erythematosus in western provns of Saudi Arabia .Rheumatol int 2011;31:23-26.

[9] Nossent JC,Swaak AJ.prevelence and significance of heamatological abnormalities in patients with systemic lupus erythematosus. QJ Med 1991;80:605.

[10] Jeffines m,Hamadeh F,Aberle T,Glenn S,Kamen DL,Kelly IA,et al Heamolytic anemia in a multi-ethnic cohort of lupus patients :a clinical and serological perspective .Lupus 2008;17:739-743.

[11] Schulze-koops H.Lymphopenia and autoimmune disease. Arthritis Res Ther 2004;6(4) 178-80.

[12] Kuwana M,Kaburaki J,Okazaki H,Ikeda Y.Two types of autoantibody-mediated thrombocytopenia in patients with systemic lupus erythematosus Rheumatology 2006;45:851-4.

[13] Schmugge M,Revel-vilk S,Hiraki,et al .Thrombocytopenia and thromboembolism in pediatric lupus erythematosus .J peditr 2003;143(5):666-9.

[14] bader-Meuner B,Armengaud JB,Haddad E Salomon R et al Initial presentyation of childhood-onset systemic lupus erythematosus: a French multicenter study.J.peditr 2005,146:648-53.

[15] bader-Meunier B,Quavtier P,Deschenes G,cochat P,haddad E,et al.Childhood-onset systemic lupus erythematosus .Arch Peditr.Feb 2003;10(2):147-157.

[16] Taddio A,Rossetto E,Rose CD,et al Prognostic impact of atypical presentation in pediatric systemic lupus erythematosus; Results from a multicenter study ,J Peditr 2010;156:972-7.

[17] Palatinus A,Adams M.Thromocytosis in systemic lupus erythematosus. Review Semin Thromb Hemost 2009;35:621-629.

[18] Leung WH,Wong KL,Lau CP,et al.Association between antiphosphlipid antibodies and cardiac abnormalities in patient with systemic lupus erythematosus. Am J Med 1990;89:411-9.

[19] Mok CC,Lau CS,Wong RWS. Risk factors for avascular bone necrosis in systemic lupus erythematosus Br,J Rheumatol 1998;37:895-900.