ejpmr, 2016,3(2), 378-380

SJIF Impact Factor 2.026

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Research Article

ISSN 2394-3211

EJPMR

STUDY OF CLINICAL FEATURES AND OUTCOME OF CHILDHOOD SYSTEMIC LUPUS ERYTHEMATOSUS

Dr. Sudhir Mehta*

MBBS, DCH, DNB (PEDIATRICS) Department of Pediatrics, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India.

***Author for Correspondence: Dr. Sudhir Mehta**

MBBS, DCH, DNB (PEDIATRICS) Department of Pediatrics, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India.

Article Received on 21/12/2015 **Article Revised on 10/01/2016** **Article Accepted on 31/01/2016**

ABSTRACT

Introduction: Systemic lupus erythematosus (SLE) is a chronic immunologic disorder with multisystem manifestations. Objective of this study was to explore clinico-laboratory manifestations and management of SLE in children. **Materials and Methods:** The study was hospital based study conducted from 2011 to 2015. Medical charts of all children and adolescent (5- 16years of age) with SLE admitted in hospital were reviewed for analysis of data. **Results:** The total number of patients was 23, with 18 girls and 5 boys. The mean age of diagnosis was 11.13±1.78. Facial puffiness and arthralgia were the commonest presentations at disease onset. The most frequent clinical features during the entire course of illness were edema (80%), anemia (78%) and fever (68%). Twenty patients underwent renal biopsy in which class IV was the commonest lupus nephritis. The commonly used drugs after prednisolone were intravenous cyclophosphamide, intravenouse methylprednisolone. Total 14 patients went into remission. One patient died due to active lupus and three due to sepsis. **Conclusion:** Lupus nephritis was the commonest feature at disease onset, at the time of diagnosis and throughout the disease course among children with SLE. The most frequently used medications were prednisolone and iv cyclophosphamide.

KEYWORDS: Childhood systemic lupus erythematosus, Lupus nephritis, ANA.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multi system inflammation.[1] Flares of the illness can involve any organ system. Diagnosis of SLE is not made until years after onset of symptoms and so alternate initial diagnoses are frequent. Making the diagnosis of SLE and ensuring appropriate early treatment may prevent devastating long-term outcomes. The clinical presentation however varies, but features such as renal, hematological and neuropsychiatric involvement may be more common in childhood SLE.[2] This article reviews common clinical features and complications of SLE in children.

MATERIALS AND METHODS

This study was done in a tertiary care hospital from 2011 to 2015. Medical records of childhood SLE cases were obtained. All patients with childhood SLE fulfilled American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus.[3] Detail clinical informations regarding mucocutaneous, cardiac, pulmonary, musculoskel et al, haematological, renal and neuropsychiatric manifesta ons etc were recorded in a proforma along with other relevant demographic profile. Laboratory data such complete blood count (CBC), urine routine, culture and 24 hour urine protein excretion, an bodies such as ANA, ds DNA, renal function tests, CXR, ultrasound abdomen etc done in each patient were recorded. Renal biopsy reports of those who underwent such procedure were also obtained. Treatments given after the diagnosis of the disease were recorded. Disease remission/progression on the basis of physical examinations and CBC, proteinuria, creatinine, ds DNA etc were also recorded as they came for follow up and medications. The investigations were performed monthly if patient has active lupus, otherwise done 3 to 6 monthly depending on disease ac vity. The duration of follow up was considered from the time of diagnosis until the patient's last hospital visit. The outcome was classified as: Remission (normal urinalysis, blood pressure, serum creatinine, no extra renal symptoms) **or** Active disease (proteinuria > 0.5g/day, microscopic hematuria >5 red cells per high power field, hypertension, extra renal manifestations) Death **or** Lost to follow-up. SPSS program 17.0 was used for statistical analysis.

RESULTS

The total number of childhood SLE was 23. Among them, female to male 18 to 5 boys. The mean age of

appearance of first symptom was 11.13±1.78 years. There was an average difference of 3-4 months from the appearance of first symptom to the diagnosis of SLE. The presenting clinical symptoms of children with SLE at disease onset are listed in **Table 1.**

The hematological with immunological findings at the time of diagnosis are shown in **Table 2**. Urine routine microscopy for protein showed that more than half of the patients had ≥ 3 + albumin. 24 hour urinary protein measurement showed nephrotic range proteinuria in 14 of the patients where as nonnephrotic range protienuria was observed in 12. Three patients had no proteinuria. Gross haematuria was seen in 13 patients. Almost half of the patients had microscopic hematuria. Granular casts was seen in two patients. 7 patients developed azotemia during disease process. Haemodialysis was performed in 5 patients.

20 patients underwent renal biopsy. The various stages of lupus nephri s (LN) based on the WHO classification on criteria for LN are as shown in the **Table 3.** More than half of the patients were in class IV. All patients received oral prednisolone at some stage of their disease. Five patients received NSAIDS) and other immunosuppressive drugs IV methyl prednisolone and cyclophosphamide were used for active disease. The number of pulse methyl prednisolone doses ranged up to 6 cycles with most patients receiving 3 cycles. Pulse iv cyclophosphamide used ranged to 12 cycles (six monthly cycle and six 3 monthly cycle).

16 developed hypertension and received various medications to lower their BP. The most common an hypertensive drug used was enalapril followed by nifedepine.

Clinical Outcomes: Follow Up

The follow up period ranged from few days to a maximum of four years. 14 patients were on remission. Longest hospital stay was 57 days and minimum 38 hours. One patient died due to active lupus and three due to sepsis.

Table1: First clinical symptom at disease onset.

Presenting first symptom	Number
Facial Puffiness	7
Joint Pain	7
Fever	5
Malar rash	2
Petechiae rash	2

Table 2: Hematological and Immunological findings at the me of diagnosis.

Laboratory data	n/N (%)
Hematological	
Anemia	68%
Thrombocytopenia	18.2%
Leucopenia	26.2%
Lymphopenia	37.4%
Immunological	
ANA	100%
Ds DNA	90%

Table 3: WHO Histopathologic findings on renal biopsy.

Lupus Nephritis Class	%
I	4%
II	18%
III	13%
IV	57%
V	8%

DISCUSSION

Childhood onset SLE is a rare disease but is reported to be comparatively severe than adults in Asians, Africans and Americans.[5,6] 23 cases were encountered in this study over the 4 years period This mean age of diagnosis is similar to other studies conducted in various centres 7,8. The relative high mean age of presentation could be largely because of under reporting or delayed referral. The female to male ratio was 18:5 which is consistent with the study done in Oman.[9] This is in contrast with study conducted by Gulay et al in Philipines and Budhathoki et al in Dharan showing female: male ratio of 10:1.[10,11] The diagnosis was based on clinical and laboratory parameters fulfilling revised criteria of 1997 American College of Rheumatology for the classifica on of SLE.[3] Since SLE has myriad systemic symptoms, differential diagnosis could be various. These figures are in accordance with lupus literature and other studies done in Asian countries.[1,9,12,13] Hematological manifestation was one of the common laboratory abnormality, which was in the form of anemia, leucopenia and thrombocytopenia. This is similar to studies conducted in Kuwait, Oman and China.[8,9,13]

The spectrum of renal involvement in different ethnici es of Asian origin are as shown in the studies-China 76.6%, Oman 64% and Kuwait-29%.[8,9,13,14] These variabilities could be because of referral patt ern, ethnic diversity etc. Rheumatological presentation was in the form of arthralgia/arthritis in 52% children which is similar to study conducted by Carian B Gulay et al in Philipino children.[10] Other presentations were alopecia, photosensi vity, mucosal ulceration, Raynaud's phenomenon, vasculitic rash and rarely discoid rash. This is in accordance with study done by Agarwal et al in Indian children and Bastug et al in Turkish children.[4,14] Serosal involvement was in the form of pericardial/pleural effusion 6 and pleural effusion 8. In

contrary to studies conducted in India and Canada, neuropsychiatric presentations are minimal in our study which evolved from 3%-15% during the clinical follow up.[15,16] Infection and active lupus were the leading causes of complication and death in childhood SLE and our observations were also similar in this study.[14]

CONCLUSION

Childhood SLE is difficult to diagnose. Patients may have varied features suggestive of SLE as well as other disease. One should suspect SLE even if classical criteria are not fulfilled and follow such patients closely.

ACKNOWLEDGEMENTS

Nil.

Funding

Nil.

Conflict of Interest

None.

REFERENCES

1. Mehta S, Sanghvi J, Kumar R. Spontaneous Pneumothorax: A rare complication of ANA negative pediatric Systemic Lupus Erythematous. Pediatric Oncall [serial online] 2015[cited 2015 April-June 1]; 12. Art #21. Available From: http://www.pediatriconcall.com/Journal/Article/Full Text.asp.
2. Livingston B, Bonner A, Pope J. Differences in clinical manifesta ons between childhood-onset lupus and adult-onset lupus: a meta-analysis. Lupus., 2011; 20(13): 1345-55.
3. Tan EM, Cohen AS, Fries JF, Masi AT, McShane DJ, Rothfield NF, et al. The 1982 revised criteria for the classifica on of systemic lupus erythematosus. Arthris Rheum., 1982; 25: 1271-77.
4. Agarwal I, Kumar TS, Ranjini K, Kirubakaran C, Danda D. Clinical Features and Outcome of Systemic Lupus Erythematosus. Indian Pediatr., 2009; 46(17): 711-15.
5. Jakes RW, Bae SC, Louthren W, Mok CC, Navarra SV, Kwon N. Systema c review of the epidemiology of systemic lupus erythematosus in the Asia-Pacific region: prevalence, incidence, clinical features and mortality. Arthritis care Res (Hoboken)., 2012; 64(2): 159-68.
6. Ramirez Gomez LA, Uribe Uribe O, Osio Uribe O, Grisales Romero H, Cardiel HM, Wojdyla D et al. Childhood systemic lupus erythematosus in La n America. The GLADEL experience in 230 children. Lupus., 2008; 17(6): 596-604.
7. Salah S, Lo y HM, Sabry SM, El Hamshary A, Taher H. Systemic lupus erythematosus in Egyp an children. Rheumatol Int., 2009; 29(12): 1463-68.
8. Alsaeid K, Kamal H, Haider MZ, Al-Enezi HM, Malaviya AN. Systemic lupus erythematosus in Kuwai children: organ system involvement and serological findings. Lupus., 2004; 13(8): 613-17.
9. Abdwani R, Rizvi SG, El-Nour I. Childhood systemic lupus erythematosus in Sultanate of Oman: demographics and clinical analysis. Lupus., 2008; 17: 83–86.
10. Gulay C B, Dans L F. Clinical presenta ons and outcomes of Filipino juvenile systemic lupus erythematosus. Pediatric Rheumatology., 2011; 9: 7.
11. Budhathoki S, Shrestha P, Bha a N, Singh R, Kalakhe B, Kumar D et al. Systemic Lupus erythematosus in childhood - a review of 11 pa ents at a single center in eastern Nepal. Nepal Medical College Journal., 2007; 9(3): 186-90.
12. Weiss JE. Pediatric Systemic Lupus Erythematosus: More Than a Posi ve An nuclear. Pediatrics in Review., 2012; 33: 62 Available from: h p:// neoreviews.aappublica ons.org/content/33/2/62.
13. Wu XY, Song HM, Zeng XF, He YY, Wang W, Li F et al. Clinical manifesta on, laboratory test, treatment and outcome of neuropsychiatric involvement in pediatric systemic lupus erythematosus pa ents. Zhonghua Er Ke Za Zhi., 2009; 47: 286–90.
14. Bastug F, Poyrazoglu H, Gunduz Z, Tulpar S, Duşunsel R. Juvenile Lupus Erythematosus: Fourteen Years of Experience. Turk J Rheumatol., 2011; 26(4): 308-15.
15. Singh S, Gupta MK, Ahluwalia J, Singh P, Malhi P. Neuropsychiatric manifesta ons and an phospholipid an bodies in pediatric onset lupus: 14 years of experience from a ter ary center of North India. Rheumatol Int., 2009; 29(12): 1455-61.
16. Benseler SM, Silverman ED. Neuropsychiatric involvement in pediatric systemic lupus erythematosus. Lupus., 2007; 16(8): 564-71.