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Systemic lupus erythematosus in paediatric age group: A rare clinical presentation

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Abstract: Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells undergo damage mediated by tissue-binding autoantibodies and immune complexes. Ninety percent of patients are women of child-bearing age. Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3-0.9 per 100.000 children and a prevalence of 3.3-8.8 per 100.000 children. In this case report we present a 4-year-old male child who presented with malar rash, oral ulceration, photosensitivity and joint pains. On histopathological examination of skin lesions, a diagnosis of lupus erythematosus was made.

Keywords: Systemic lupus erythematosus, paediatric age group, autoimmune diseases, autoantibodies.

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells undergo damage mediated by tissue-binding autoantibodies and immune complexes [1]. Ninety percent of patients are women of child-bearing age. Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3-0.9 per 100.000 children-years and a prevalence of 3.3-8.8 per 100.000 children [2]. In this case the patient was a 4 year old male child which is a rare presentation. The recent used diagnostic criteria widely used for SLE is 2015 ACR/SLICC Revised criteria for diagnosis of Systemic Lupus Erythematosus which is based on point systems. Patient with 4 points out of 16 have definite diagnosis of SLE, with 3 points highly suggestive of SLE, with 2 points probable SLE and with one point possible SLE are the diagnosis [3].

CASE SUMMARY

A 4 year old male child presented to the dermatology outpatient service of Guru Gobind Singh Medical College and Hospital, Faridkot with complaints of oral ulceration, malar rash, photosensitivity and joint pains. History dates back to 3 years ago when child had chief complaints of erythematous patch on face, reddish spots on scalp which were followed by oral ulcerations, one week after the onset of symptoms. The patient was getting treatment from a nursing home in Rajasthan where he was put on Tab Prednisone and Hydrocortisone cream and was asked to follow up after 4 months but was not relieved of these symptoms. The patient then switched to another private practitioner who also gave him steroids but no relief. After one year of no relief he presented to dermatology outpatient service of Guru Gobind Singh Medical College and Hospital, Faridkot. On examination erythematous patch on face, reddish spots on scalp and oral ulcerations were seen.

The blood sample of the patient was taken for complete blood count (CBC) and Antinuclear antibody profile (ANA). The ANA profile was derranged (Table no.1). The child was put on Tab Prednisone and Hydrocortisone cream and a skin biopsy was performed and sample was sent pathology department for histopathological evaluation. Unfortunately the child didnot survive.

The histopathological examination revealed thinning and ulceration of epidermis along with basal cell layer vacuolation. (Fig.4,5) Also the dermis showed presence of moderate periappendageal and perivascular lymphocytic infiltrate thus suggesting the diagnosis of Lupus erythematosus which was confirmed on immunofluoroscence.

Table-1: ANA WESTERN BLOT PROFILE

TEST	RESULT	NORMAL
ANA(Antinuclear antibodies)	97IU/ml	Negative<40 IU/ml Positive>45 IU/ml
dsDNA(Anti double stranded DNA)	116 IU/ml	Negative<50 IU/ml Positive>50 IU/ml
SS-A(Anti sjogren syndrome related antigen A)	72 IU/ml	Less than 30 IU/ml
SS-B(Anti sjogren syndrome related antigen B)	34 IU/ml	Less than 30 IU/ml
Anti-SCL-70(Anti topoisomeraseI)	36 IU/ml	Less than 30 IU/ml
Anti-SM(Anti smith)	76 IU/ml	Negative< 40 IU/ml Moderately positive 40-45 IU/ml Highly positive>45IU/ml
ANTI U1 RNP(Anti U1 ribonucleoprotein)	25 IU/ml	Less than 20 IU/ml

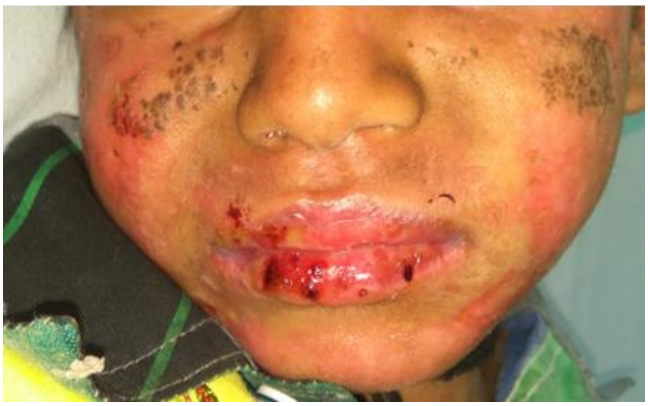

Fig-1: Child had erythematous rash on face and oral ulcerations

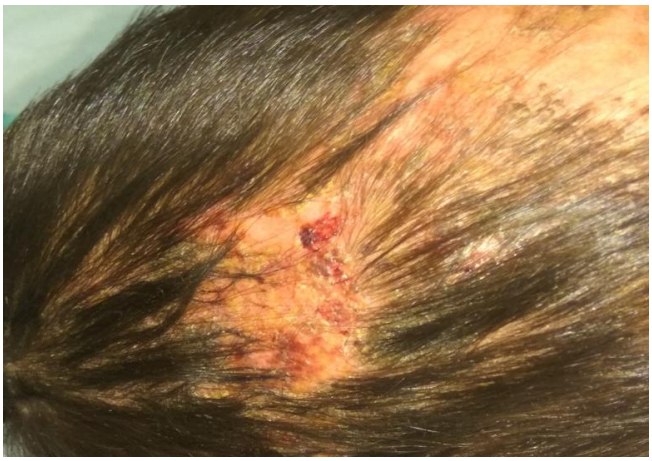

Fig-2: Photograph of child showing reddish spots on scalp

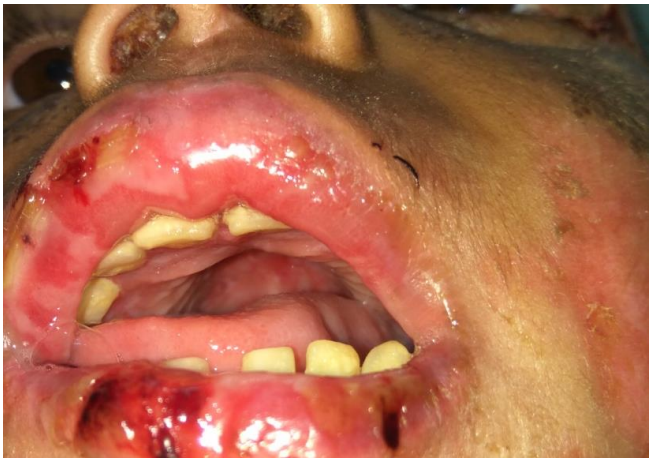

Fig-3: Photograph of child showing oral ulcerations

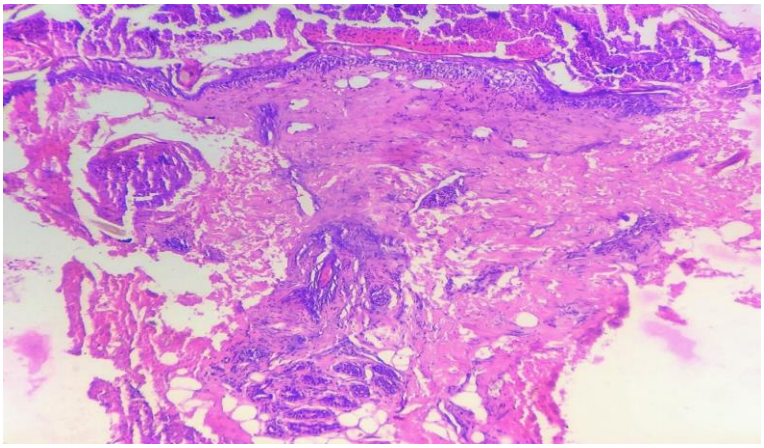

Fig-4: Photomicrograph showing thinning and ulceration of epidermis with periappandageal and perivascular lymphocytic infiltrate (100x)

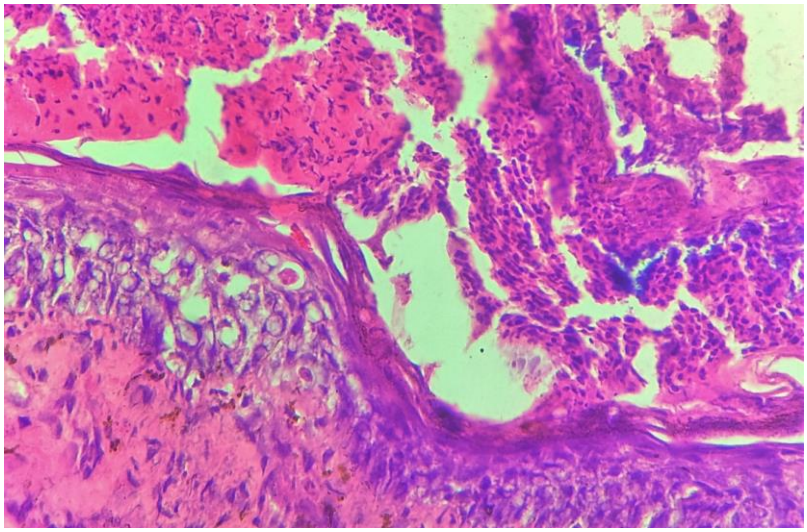

Fig-5: Epidermal lining exhibiting basal cell layer vacuolation, epidermal thinning and ulceration

DISCUSSION

SLE is more common in female of child bearing age group where the reported female: male ratio is 8-15:1 [4]. Paediatric SLE is a rare disorder with very few cases described in age group less than 2 years. Childhood-onset SLE is a lifelong autoimmune disease that may be difficult to diagnose due to its multisystem involvement, and heterogeneity of clinical manifestations. Most studies report a median age of onset of cSLE between 11-12 years; the disease is quite rare under the age of 4 years while in this patient the age was 04 years. As in adult onset SLE, approximately

80% of patients with cSLE are female [2,5] but in this case the patient was a male which is also a rare presentation.

The recent used diagnostic criteria widely used for SLE is 2015 ACR/SLICC Revised criteria for diagnosis of Systemic Lupus Erythematosus which is based on point systems. Patient with 4 points out of 16 have definite diagnosis of SLE, with 3 points highly suggestive of SLE, with 2 points probable SLE and with one point possible SLE are the diagnosis [3].

It follows a more aggressive disease course than adult-onset SLE, with greater disease activity at presentation and over time, and consequently leads to greater morbidity and mortality than adult-onset SLE [6]. Diagnosis of SLE can be confirmed by histopathological and direct immunofloroscence examination of skin biopsy specimens of an active lesion. Patient had histopathological features of SLE.

CONCLUSION

Although SLE is primarily a disease of adults it can present itself in pediatric age group as well and although it is much more common in females it can present in males as well as highlighted in this case report.

REFERENCES

1. Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18e. New York, NY:Mc Graw-Hill; 2012
2. Hiraki LT, Benseler SM, Tyrrell PN, et al. Ethnic differences in pediatric systemic lupus erythematosus. Journal of Rheumatology. 2009; 36:2539–46
3. Salehi-Abari I. ACR/SLICC Revised Criteria for Diagnosis of Systemic Lupus Erythematosus. Autoimmune Dis Ther Approaches Open Access. 2015;2:114.
4. Lu LJ, Wallace DJ, Ishimori ML, Scofield RH, Weisman MH. Review: Male systemic lupus erythematosus: a review of sex disparities in this disease. Lupus. 2010 Feb 1;19(2):119-29.
5. Pluchinotta FR, Schiavo B, Vittadello F, Martini G, Perilongo G, Zulian F. Distinctive clinical features of pediatric systemic lupus erythematosus in three different age classes. Lupus. 2007 Aug;16(8):550-5.
6. Levy DM, Kamphuis S. Systemic lupus erythematosus in children and adolescents. Pediatric Clinics of North America. 2012 Apr 30;59(2):345-64.