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SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) – A RARE CASE REPORT

Paediatrics

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ABSTRACT

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and presence of circulating auto-antibodies directed against self-antigens.We herein, report a case of a 8-year old female child diagnosed as Systemic Lupus Erythematosus due to its rare occurrence in pediatric age group.

KEYWORDS

INTRODUCTION

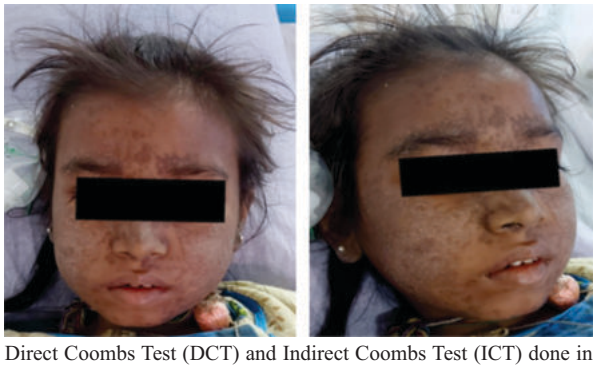
Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and presence of circulating auto-antibodies directed against self-antigens. Although, nearly every organ maybe affected, most commonly involved are the skin, joints, kidneys, blood forming cells, blood vessels and central nervous system.[1] The occurrence of Systemic Lupus Erythematosus (SLE) in children is very rare. About 20% of all cases are diagnosed in the first two decades of life.[2] The childhood onset of SLE occurs between the ages 3 and 15, with girls outnumbering boys in the ratio 4:1.[4]SLE is difficult to diagnose as it mimics other diseases.

Case Report

A 8-year old girl was admitted in New Civil Hospital in Surat with the complaint of high grade fever, and cough since 4 days with history of rash 15 days before admission.

On general examination of the patient, hyperpigmented plaque with scaling present over malar area of face, Ala of nose, forehead, ears with sparing of nasolabial folds. Hyperpigmented plaque with scaling present over the abdomen, upper back, upper part of buttocks, over b/l upper and lower limbs. Single ulcer with crusting present over upper ababdomen. Sparse hair which is Positive for hair pull Test suggestive of Telogen Effluvium.

Routine investigations were sent in which hemoglobin was found to be 5.2g/dl. Further investigation for anemia shows high reticulocyte count(6%), raised S.ferritin (1485 ng/ml), ESR=140mm, raised S.LDH (851mg/dl), S.Uric acid 4.6mg/dl. Sickling test done for patient with family screening and all three came out negative.



Direct Coombs Test (DCT) and Indirect Coombs Test (ICT) done in which antibody screening and antibody identification suggestive of Pan Positivity. Ultrasonography abdomen, pelvis and thorax done suggestive of mild pleural effusion. 2D-Echo suggestive of mild pericardial effusion. A possibility of Systemic Lupus Erythematosus was being considered for this patient for which ANA study was done by Immunoflorescence and suggestive of raised ANA titre 1:320.

DISCUSSION

The diagnosis of SLE can be established through total score of clinical domains and immunological domains >=10 and >=1 clinical criteria and ANA titre >= 1:80 on Hep-2 cells required to classify SLE. [3]

In our case study, clinical domains and criteria such as autoimmune hemolysis (4), non scaring alopecia(2), pleural/pericardial effusion (5), fever(2), which results in a total score of 13, with ANA titre 1:320.

The disease severity varies from mild to severe, and requires long term and often aggressive treatment.

CONCLUSION

Since Systemic Lupus Erythematosus is a rare multisystem inflammatory disorder that is difficult to diagnose especially in children because of its rare occurrence, the clinician must be aware of the various possible presentations of the disease and systemic complications in children in order to identify and establish the diagnose of SLE, so that timely and effective management can be initiated. Long term follow up of patient and appropriate changes in management according to disease activity is required so that we can reduce mortality and morbidity that arises due to SLE.

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