

Childhood Systemic Lupus Erythematosus; A Rare Multisystem Life-threatening Disorder: a case of 13 years-old-girl with oral involvement and joint pain as primary clinical manifestation

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune illness that affects several organs, and the most frequent renal consequence of SLE is lupus nephritis (LN). It is a rare, systemic disease, possibly fatal autoimmune illness with high morbidity. SLE prevalence has been observed to range between 14 and 60 per 100,000 people. A 13-year-old female child was admitted to the hospital in the paediatric ward with the chief complaint of skin lesion over sun-exposed and bleeding from oral mucosa. Blood samples were sent to the laboratory for investigations, and blood cultures were also sent to the microbiology lab. As soon as she was admitted, treatment was started without any delay. Blood investigations show a decreased level of WBC count, i.e. 3500 cu/mm, and the blood culture revealed the organism's growth. Hence patient was started on injectable antibiotics. After 15 days of a continuous course of antibiotics, the patient started recovery. The rashes started disappearing a little bit. Physiotherapy is begun for a child with joint pain where she experiences relief.

Keywords: Systemic lupus erythematosus (SLE), autoimmune illness, skin lesion, mucosal bleeding, physiotherapy

INTRODUCTION

Childhood-onset systemic lupus erythematosus (cSLE) is a rare, systemic disease, possibly fatal autoimmune illness with high morbidity. There are few evidence-based guidelines and care frequently relies on clinical experience. (1) In a population survey of 91,888 people, three cases of SLE were found, yielding a point prevalence of 3.2 per 1 lakh (95 percent CI = 0 to 6.86 per 1 lakh). SLE prevalence has been observed to range between 14 and 60 per 100,000 people. As a result, the prevalence rate of SLE in India is relatively low. (2) Oral ulcers are most prevalent mucosal manifestation in juvenile (SLE) patients. Mouth ulceration are a crucial clinical characteristic; nevertheless, the nomenclature of mucosal ulcers, particularly in Juvenile SLE patients, is sometimes unclear and not well-defined.

Numerous clinical presentations of oral ulcers in Juvenile SLE, and certain lesions appear while the disease is progressive, indicating that the condition should be managed as soon as possible. (3)

Appropriately 15 percent of people with SLE may experience illness in infancy or adolescence. Because of the wide variety of probable clinical symptoms of SLE, a diagnosis in a general paediatric or community environment may be challenging.

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Access this article online

Quick Response Code:



Website:

www.pnrjournal.com

DOI:

10.47750/pnr.2022.13.03.144

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How to cite this article: Namrata Agase, Ruchira Ankar, Tejaswee Lohakare, Roshan Umate, Childhood Systemic Lupus Erythematosus; A Rare Multisystem Life-threatening Disorder: a case of 13 years-old-girl with oral involvement and joint pain as primary clinical manifestation, J PHARM NEGATIVE RESULTS 2022;13: 934-937.

Fever, exhaustion, weight loss, arthritis, rash, and kidney failure are frequent symptoms of SLE in paediatric children and early adolescents. (4) SLE can cause significant morbidity in children and adolescents. (5)

SLE is an autoimmune illness that affects several organs, and the most frequent renal consequence of SLE is lupus nephritis (LN). Belimumab is a completely humanised monoclonal antibody that can lower the number of B cells in the body, decreasing the generation of autoantibodies. Belimumab can enhance the SLE response index and disease activity score in both adults and children, as well as postpone the progression of LN, and so plays a vital role in the treatment of SLE and LN.(6) Children with SLE suffer from chronic disease and commonly feel discomfort as a result of their condition. They may also experience social isolation.(7)

CASE REPORT

A 13-year-female child was admitted to the hospital in the paediatric ward with the chief complaint of skin lesion over sun-exposed areas and bleeding from oral mucosa. As narrated by her mother-child was alright three months before; she started developing pain in knee joints, elbow and wrist joints and developed skin lesions over the palm of soles and sun-exposed areas. On physical examination, it is observed that the patient has itching, oral mucosal bleeding, joint pain, high-grade fever, intermittent and not associated with chills or rigours, and history of photosensitivity. Blood samples were sent to the laboratory for investigations, and blood cultures were also sent to the microbiology lab. As soon as she was admitted, treatment was started without any delay. Paediatrician has prescribed Inj. Rituximab 375 mg once a day, tab. Hydroxychloroquine 100 mg twice a day, Tab. Omnacortil 10 mg once a day for 15 days. Inj. Ceftriaxone 1 gm twice a day, Inj. Pantoprazole 20 mg once a day and Tab. Paracetamol 650 mg ½ tab whenever necessary in case of fever. (fig 1)



Figure 1: image showing marks on feet

Blood investigations show a decreased level of WBC count,

i.e. 3500 cu/mm, and the blood culture revealed the organism's growth. Hence patient was started on injectable antibiotics. After 15 days of a continuous course of antibiotics, the patient started recovery. The rashes started disappearing a little bit. Physiotherapy is begun for a child with joint pain where she experiences relief. Physiotherapy has an excellent benefit for her. On the 18th day, the child was planned to be discharged on oral antibiotics.

DISCUSSION

According to a study, SLE is a chronic neurodegenerative condition that can be tough to diagnose due to the involvement of several systems and the wide range of clinical symptoms. It has more rapid onset than adult SLE, as time over progression and presentation also increases with time that leads to high mortality and morbidity rate. Furthermore, children must cope up with this unpredictable, and relapsing disease throughout adolescence, with the help of physical appearance self esteem and identity also growing, and improve overestimation of self decision – making and responsibility for their health is prevalent. These cSLE specifically recognize the issue which are crucial for optimising management of SLE in children. There are few long-term outcome studies of cSLE; nevertheless, new research is being performed to explore adult-aged cSLE cohorts. These findings will aid in better predicting the long-term prognosis of cSLE and may serve as the cornerstone for a more tailored management and therapy approach for children and adolescents with cSLE.(8-19)

A Charras, in his study, says that genetic factors have a crucial part in the pathogenesis of jSLE since single-gene mutations cause illness in more than 7% of individuals. The remaining patients have genetic variations required for illness development but need other variables to be present. Increased 'genetic influence' is expected to contribute to illness development and severity. Epigenetic events, which have recently been studied in jSLE, add to the number of pathogenic pathways that might serve as biomarkers and therapeutic targets. Age-specific categorisation criteria and therapy objectives must be specified to allow relevant and patient-oriented pediatric research, as currently available methods established for adult-onset SLE have limitations in the pediatric group. Understanding the pathophysiology of jSLE has advanced significantly. Only age-specific techniques, categorisation criteria, and treatment objectives may be used to conduct meaningful laboratory and clinical research. (20-33)

CONCLUSION:

A diagnosis of systemic lupus erythematosus in a general paediatric or community environment may be challenging. Here, in this case of SLE, with the help of medical treatment that is antibiotics and steroids patient has started her recovery;

her prognosis was good and hence discharged on oral antibiotics.

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