ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 04 | April - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CHILDHOOD SYSTEMIC LUPUS ERYTHEMATOSUS WITH VASCULITIS- A CASE REPORT

Dermatology

Dr. Krithi Subhas Chandra* MBBS. Resident in Department of Dermatology, Venereology and Leprosy, Kempegowda Institute of Medical Sciences, Bangalore.*Corresponding Author

Dr. Sharath Kumar B.C MBBS, MD, DNB. Professor and Head of Department, Department of Dermatology, Venereology and Leprosy, Kempegowda Institute of Medical Sciences, Bangalore.

ABSTRACT

Systemic Lupus Erythematosus (SLE) is an autoimmune connective tissue disorder with varied systemic manifestation. We report a 7 year old girl who presented to us with a history of redness and raised lesions over the face for 7 months, which were gradually progressive and increased on exposure to sunlight. On examination, we found multiple ill-defined erythematous scaly plaques over the forehead, bilateral malar region and nose with involvement of the concha and scalp. Carpet tack sign was present. Oral cavity examination revealed ulcers over the hard palate. She also had multiple erythematous crusted papules over bilateral lower limbs, arms and trunk. Specific tests showed elevated ANA titres, positive anti dsDNA antibodies and anti Smith antibodies. Urine routine showed proteinuria and elevated protein creatinine ratio. She was diagnosed to have childhood SLE, which was confirmed on histopathological examination; with associated small vessel vasculitis. The child was treated with intravenous Methylprednisolone for 3 days followed by oral steroids and Oral Hydroxychloroquine sulphate. Topically, she was given potent corticosteroids and sunscreen. After 1 month of initiating treatment, the skin lesions were found to have subsided, with presence of hyperpigmentation and minimal scarring. Our initial treatment was found to be effective in preventing further renal impairment, with a decrease in proteinuria and normalisation of protein creatinine ratio. We concluded that though childhood SLE is a very rare occurrence, it should be kept in mind when encountering a child with scaly plaques in a photo distributed area. It is a challenging condition to treat and requires a multi-disciplinary approach to management, with a need for regular monitoring and counselling. We found that the early aggressive treatment of the condition can prevent the progression to permanent renal damage.

KEYWORDS

Systemic Lupus Erythematosus, Lupus Nephritis, Childhood Sle, Vasculitis.

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is an autoimmune connective tissue disorder with varied systemic manifestation. SLE is very rare among children, with it being reported in less than 3 per 1,00,000 children [1, 2]. The age of onset is usually 3-15 years [3], however cases have been reported in infants as young as 5 months of age [4]. It is found to be more common in girls than boys in the ratio of 4:1 [3]. Childhood onset Discoid lupus erythematosus has a higher chance of developing systemic disease. Childhood SLE is associated with significantly higher morbidity and mortality than adult SLE due to increased likelihood of underlying organ involvement, of which nephritis is the most common [5]. Treatment modalities for childhood lupus that have been suggested include corticosteroids, hydroxychloroquine, immunosuppressants, intravenous immunoglobulins, plasma exchange, and biologicals [5].

Case report

A 7 year old girl presented to us with a history of redness and raised lesions over the face for the past 7 months, which were gradually progressive. They were found to increase on exposure to sunlight. She had history of intermittent fever which decreased with medication. She also developed painful oral ulcers due to which her food intake had decreased. There was no history suggestive of Raynaud's phenomenon, joint pain or neuropsychiatric symptoms.

On examination, pallor was present. Her developmental milestones were appropriate for her age. Cutaneous examination revealed multiple ill-defined erythematous scaly plaques over the forehead, bilateral malar region and nose with involvement of the concha (*figure 1*). Multiple alopecic plaques with scaling were present on the scalp. Carpet tack sign was present. Oral cavity examination revealed ulcers over the hard palate (*figure 2*). She also had multiple erythematous crusted papules over bilateral lower limbs, arms and trunk (*figure 3*). Nail fold capillaroscopy was normal.

Figure 1: (a) Multiple erythematous scaly plaques over the nose, bilateral malar region, nose, and forehead. (b) Scaly erythematous plaque over the concha (Shuster's sign)

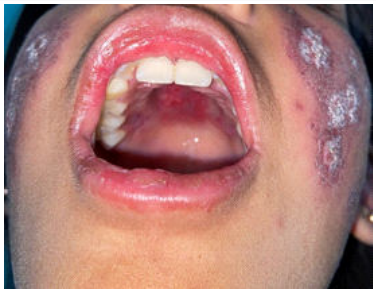

Figure 2: Erosions present over the hard palate and soft palate.

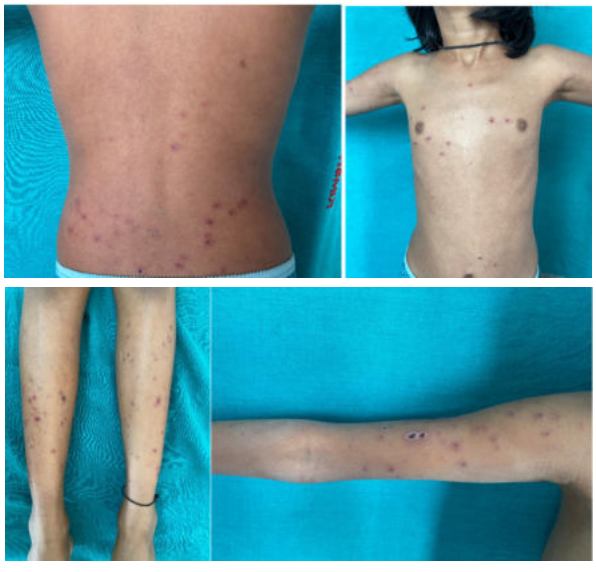

Figure 3: Multiple erythematous non blanching purpuric macules with few crusted lesions present over the trunk (a, b), bilateral legs and arms (c, d).

Blood investigations revealed pancytopenia. ESR and CRP were elevated. Specific tests showed elevated ANA titres, positive anti

dsDNA antibodies and anti Smith antibodies. Liver enzymes were raised. Urine routine examination showed proteinuria. Protein creatinine ratio was found to be elevated. Chest X-ray and ophthalmologic tests were also normal. The histopathological examination of skin biopsy specimen showed hyperkeratosis, focal parakeratosis of the epidermis with thinning, basal vacuolar degeneration with melanin incontinence. Few civatte bodies were seen. Papillary dermis showed superficial perivascular and interstitial infiltrate predominantly of neutrophils and lymphocytes (*figure 4*).

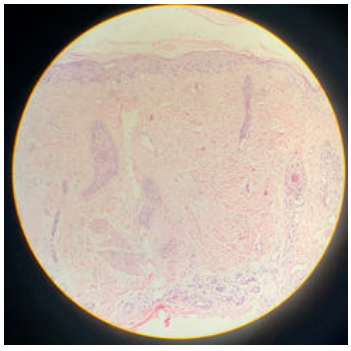

Figure 4: Histopathology showing hyperkeratosis, focal parakeratosis of the epidermis with thinning, basal vacuolar degeneration. Papillary dermis showed superficial perivascular and interstitial infiltrate predominantly of neutrophils and lymphocytes

On the basis of clinical presentation, laboratory findings and histopathology, diagnosis of childhood-onset SLE was done. The presence of multiple crusted papules in the bilateral lower limbs was suggestive of the presence of cutaneous small vessel vasculitis also.

The child was treated with systemic corticosteroids (Intravenous Methylprednisolone for 3 days followed by oral Prednisolone 1mg/kg/day) and Hydroxychloroquine sulphate (5mg/kg/day), potent topical corticosteroids. The parents were counselled regarding strict photoprotection and the need for regular follow up and monitoring of renal function. Sunscreen and Vitamin D supplements were also given. After 1 month of initiating treatment, the skin lesions were found to have subsided, with presence of hyperpigmentation and minimal scarring.

DISCUSSION

SLE is an autoimmune condition, the exact etiology of which is unknown. It is proposed to be due to a complex interaction between genetic and environmental factors leading to the production of autoantibodies, eventually resulting in organ damage [6]. SLE is diagnosed based on the American College of Rheumatology and European League against Rheumatism Criteria [7]. There is no separate criteria for childhood SLE. Our case met the entry criteria of ANA titres more than 1:80, and had a score of more than 10 from the additional criteria based on clinical, histopathology, and serological tests. Small vessel vasculitis is an atypical manifestation of SLE found in 11-36% cases [8]. In our case we also found the presence of multiple, non-blanching, crusted, purpuric lesions which was diagnosed clinically as cutaneous small vessel vasculitis, and was confirmed by histopathology.

Our case also showed positive double stranded DNA, which signifies the considerable risk of developing lupus nephritis. [9]. Our case was found to have proteinuria and elevated urine protein creatinine ratio, indicating the presence of renal impairment. Early aggressive management of SLE has been recommended to prevent the progression to permanent renal damage. Our initial treatment was found to be effective in preventing further renal impairment, with a decrease in proteinuria and normalisation of protein creatinine ratio.

Corticosteroids are the first line treatment in childhood SLE. Non-steroidal immunosuppressants such as Azathioprine and Mycophenolate Mofetil have also been given. In our patient, we administered Intravenous Methylprednisolone for 3 days followed by oral steroids at the dose of 1mg/kg/day. Oral Hydroxychloroquine sulphate has been used in children with extensive cutaneous involvement and complicated with arthritis. Our patient was given Hydroxychloroquine sulphate at the dose of 5mg/kg/day. Refractory cases have been treated with Rituximab, a monoclonal anti-CD20 antibody at the dosage of 750mg/m2, administered intravenously at an interval of 2 weeks. Children with SLE on systemic therapy need clinical evaluation every 4-6 months along with repetition of laboratory parameters every 6 months [5].

CONCLUSION

Childhood SLE is a rare occurrence, which should be kept in mind when encountering a child with scaly plaques in a photo distributed area. It is a challenging condition to treat and requires a multi-disciplinary approach to management, with a need for regular monitoring and counselling. We found that the early aggressive treatment of the condition can prevent the progression to permanent renal damage.

Conflict of Interest

None

Consent

Informed consent has been obtained from the parents of the child for this publication.

REFERENCES

1. Chandrasekaran, A. N., Rajendran, C. P., Ramakrishnan, S., Madhavan, R., & Parthiban, M. (1994). Childhood systemic lupus erythematosus in south India. *The Indian Journal of Pediatrics*, *61*(3), 223-229.
2. Ali, U. S., Dalvi, R. B., Merchant, R. H., Mehta, K. P., Chablani, A. T., Badakere, S. S., & Bhatia, H. M. (1989). Systemic lupus erythematosus in Indian children. *Indian pediatrics*, *26*(9), 868-873.
3. Kumar, A., Kumar, P., Bhengra, M. P., Kumar, P., & Chaudhary, S. S. (2016). Childhood-onset systemic lupus erythematosus–A case report. *Journal of Pakistan Association of Dermatology*, *26*(1), 76-79.
4. Dogra, D., & Khanna, N. (1996). Systemic lupus erythematosus in an infant. *Indian pediatrics*, *33*, 779-780.
5. Palit, A., & Inamadar, A. C. (2012). Current treatment strategies: collagen vascular diseases in children. *Indian journal of dermatology*, *57*(6), 449.
6. Vaillant, A. A. J., Goyal, A., Bansal, P., & Varacallo, M. (2020). Lupus Erythematosus. *StatPearls [Internet]*.
7. Aringer, M., Costenbader, K., Daikh, D., Brinks, R., Mosca, M., Ramsey.Goldman, R., ... & Johnson, S. R. (2019). 2019 European League Against Rheumatism/American College of Rheumatology classification criteria for systemic lupus erythematosus. *Arthritis & rheumatology*, *71*(9), 1400-1412.
8. Barile-Fabris, L., Hernández-Cabrera, M. F., & Barragan-Garfias, J. A. (2014). Vasculitis in systemic lupus erythematosus. *Current rheumatology reports*, *16*(9), 440.
9. Bolognia J, Schaffer J, Cerroni L. (2018). *Dermatology.*4th edition. Elsevier. p663.