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CASE REPORT

A Case of Infantile Systemic Lupus Erythmatosis (iSLE) - Sharing the Experience

BUSHRA RAFIQUE

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Author's affiliations

Correspondence to:

Bushra Rafique,
Department of Child Health,
Royal Hospital, Muscat,
Sultanate of Oman

E-Mail:
bushra.nadeem@yahoo.com

ABSTRACT

A 3 month old Omani boy developed fever with skin rash. He was taken to many local health centers, got treatment but without any lasting benefit. At the age of eight months he developed status epilepticus and was brought to Royal Hospital, Muscat, Oman, for further management. He was thoroughly investigated and diagnosed as infantile Systemic Lupus Erythmatosis (iSLE). He responded very well to the treatment and is now in complete remission.

iSLE is a rare disorder; this made us to search and review the experience of the experts worldwide.

Key words: Pediatric SLE, subtypes, management, outcome

INTRODUCTION

SLE is an autoimmune disease of unknown origin. When disease presents below 16 years of age it is called Pediatrics SLE (pSLE) which constitutes 10-15% of cases[1]. It is further divided into three groups:

1. Infantile SLE (iSLE): when disease presents below two years of age.
2. Pre-pubertal SLE: is diagnosed between 2-10 years of age.
3. Post-pubertal or juvenile SLE (jSLE): presents between 11-16 years.

iSLE is the smallest among these three groups[2]. This was the stimulus to review the literature and share our experience.

CASE HISTORY

An eight month old Omani boy, weighing eight Kg, was shifted from a peripheral hospital because of uncontrolled seizures. He had multiple skin lesions and oral ulcers since 3 month of age. He also had fever off and on since that time. Parents consulted local health centers whenever he was sick. Different treatments, including antibiotics were given but response was partial and temporary. Since last three days before presentation to us he was running high grade fever and then developed generalised tonic clonic seizures. He was given diazepam and then loaded with phenytoin. As he continued to have fits, so was referred to Royal Hospital Muscat, where he was directly shifted to Pediatric Intensive Care Unit (PICU), intubated, ventilated and paralysed.

Past history: He had BCG related axillary abscess at three months of age, which needed incision and drainage.

Family history: He was the only child, born nine years after marriage. No family history of any autoimmune or other significant disease.

On examination: On presentation he was a sick looking child with multiple erythematous annular lesions all over the body especially on the face, ear, abdomen and genitalia. His head was full of dandruff and there were many mouth ulcers. Liver was 2 cm, while tip of spleen was palpable below the costal margin. Cardiovascular and respiratory systems were unremarkable. Developmentally he was upto the age. There was no CNS deficit.

Investigations: Hemoglobin – 5 gm/dl, white blood cells – $2.9x10^9$/L, platelets – $227x10^9$/L, reticulocytes 0.5% Blood film: aniso-poikilocytosis, elliptocytosis, tear drop cells, occasional fragmented RBCs, mild toxic granulation and vaculation. C-Reactive Protein(CRP) – 9 mg/L, Erythrocyte Sedimentation Rate (ESR) – 81mm/1st hour. Liver Function Tests (LFT): Bilirubin – 6 mmol/L, Alanine Aminotransferase (ALT) – 43 iu/L, Alkaline Phosphatase (ALP) – 50 units/L, Protein – 59 g/L, Albumin – 25 g/L Renal profile, bone profile. Hemoglobin electrophoresis, Uric acid, lipid profile, Immunoglobulin level – normal Coagulation profile: Prothrombin Time (PT): 10.2 seconds – control: 10 seconds, Activated Partial Thromboplastin Time (APTT): 48.7 seconds – control 35 seconds, Fibrinogen: 2.93 gm/L – normal: 2-4 gm/L, Thrombin Time (TT): 21.4 seconds – control 12 seconds. Complements: C1q – 124 mg/dl (100-125mg/dl), C3 – 1328 mg/dl (790-1520mg/dl), C4 – 588 mg/dl (160-380mg/dl), CH50 - <10 mg/dl(37-66mg/dl) Ferritin – 3318 mcg/L (7-140 mcg/L), Direct Coomb's Test (DCT) –ve, Serum Ammonia (NH4) – 69 mmol/L (10-45 mmol/L), Lactate – 1.5 mmol/L (0.5-2 mmol/L), Lactate Dehydrogenate (LDH) – 941 iu/L (95-190 iu/L) Blood culture x 5 times – once positive for Salmonella specie, CSF culture x 3 times – once positive for Salmonella specie, Urine c/s –ve, Human Immunodeficiency Virus (HIV) –ve, Fluorescent Treponemal Antibody Absorption (FTA) –ve, Lymphocyte subsets – generalised lymphopenia with normal CD4:8 ratio, Immunodeficiency work up – normal, Anti-Nuclear Antibodies (ANA) +ve 1:320, Anti-double standard Deoxyribonucleic Acid (dsDNA) –ve, Anti-nuclear Cytoplasmic Antigen (ANCA) –ve, Extractable Nuclear Antigen (ENA): Anti Ribo-nucleoprotein (Anti RNP) –ve, Anti SCl-70 –ve, Anti sm – weak+, Anti-SSA(Ro)+ve, Anti-SSB(La)-ve, Anti-Jo1 –ve, Proliferating Cell Nuclear Antigen – ve Lupus Antigen Screen: PTT – lupus sensitive 31.3 seconds (23.9-34.9), Lupus screen 1 – 57.7 seconds (31-44), Lupus screen 2 – 37 seconds, Lupus confirm – moderate+ 1.6 (0.1-1.1), Chest x-ray – normal, ultra sound abdomen – mild hepatosplenomegaly, CT head – ventriculo-megaly, non-communicating hydrocephalus, generalised brain atrophy, left parietal brain is edematous with abnormal increased gyral enhancement - ? Vasculitis MRI brain – subacute left subdural hematoma. Mother's Anti-nuclear antibodies (ANA) and Extractable Nuclear Antigen Antibodies (ENA) –ve.

Management: The child presented to us with resistant seizures, so he was intubated, ventilated and paralysed. Phenytoin was continued to control the fits. Ceftriaxone and Meropenem were given as Salmonella was sensitive to these drugs and child showed good response. On this regimen his fits were controlled, he gained consciousness and fever subsided. However skin lesions and irritability remained the same. Chlorpheneramine was given to control itching till results of investigations were available. Once diagnosis of infantile SLE was confirmed he was started on following regimen.

1. Mycophenolate Mofetil (syrup), 125 mg, BD.
2. Hydroxychloroquine (syrup), 50 mg, OD.
3. Methyl prednisolone, 30 mg/kg/dose in 100 ml of pediatric saline over 2 hours under cardiac monitor for 3 consecutive days.
4. Followed by Prednisolone 1 mg/kg/day for 1 month and then half of this dose for one more month.
5. Betamethasone cream for local application on the skin lesions.
6. Calcium gluconate, 250 mg, OD.
7. Ferrous sulphate 20 mg, OD.
8. ` Alfa calcidol 1 mcg, OD.
9. No live vaccine till on medications.
10. He was given varicella Ig and acyclovir when he developed chicken pox.

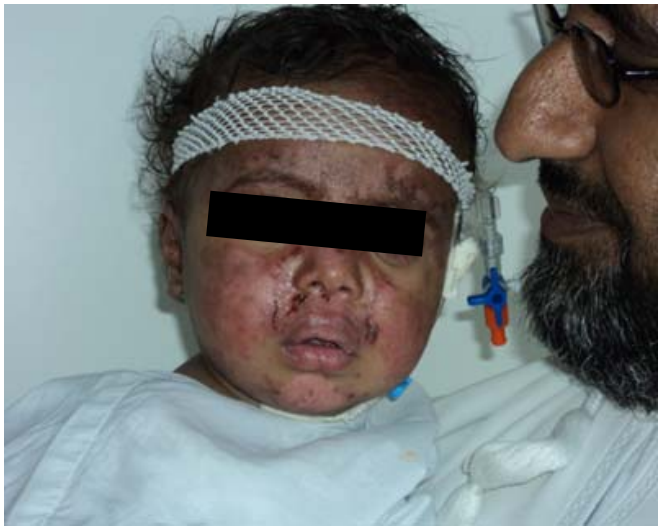

Fig 1: Patient at presentation

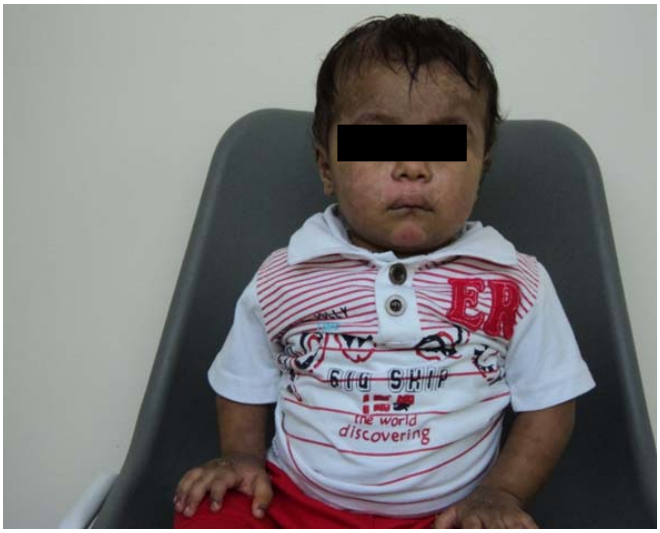

Fig 2: Patient after treatment

DISCUSSION

Systemic Lupus Erythmatosis (SLE) is an autoimmune disease of unknown origin. Autoantibodies are most of the time directed against joints, kidneys, hematopoietic cells and central nervous system etc.

SLE is a disease of adolescent age, however 10-15% of the cases present before 16 years of age[1]. This pediatric population is further divided into three groups depending upon the age of presentation. This grouping is important because of different gender predisposition, organ involvement, severity, morbidity and mortality.

Infantile SLE (iSLE) is the least common group. Both sexes are affected equally. Common presentation is with fever, rash and irritability[5]. History is short and child looks miserable. Hematological findings are also different from adolescent group. Older patients present with cytopenia while infants presents with leukocytosis[6]. However our patient presented with bi-cytopenia. Diagnostic tests remain the same like high ESR and positive auto-antibodies especially ANA, anti dsDNA, Anti-Sm, Anti-cardiolipin (ACL) and Lupus Anti- Coagulants (LAC).

The disease presents acutely, run severe course and demands early introduction of immunosuppressive treatment along with steroids. Morbidity and mortality remains high in this age group. Survival is 69.5% at 1 year, 61.5% at 3 years. While survival rate in jSLE is 100% at 1 year and 86% at 10 years[7].

Infantile SLE – iSLE should not be confused with Neonatal SLE – NLE. This is seen in children born to mothers with SLE. Trans-placental passage of Anti-Ro/SSA and anti-La/SSB produces discoid rash, cytopenia and congenital heart block. Skin and blood pathology reverses around six months of age while heart block may be permanent. It is important to keep iSLE in the differential diagnosis when a child presents with fever, rash and irritability. Early treatment can induce remission and decrease morbidity and mortality.

CONCLUSION

Infantile SLE (iSLE) is a rare subtype of pediatric SLE (pSLE). Fever, rash and irritability in an infant should raise the suspicion. The onset of disease is acute and it runs an aggressive course. Proper investigations can clinch the diagnosis. Specific treatment and regular follow up can help to control the disease. Literature review however revealed guarded prognosis.

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