

Systemic Lupus Erythematosus with Multiple Faces in a Girl of 11 Years

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Abstract

Case Report

Systemic lupus erythematosus in children (childhood onset SLE/cSLE) have a great variability in disease presentation. Any organ system can be involved in cSLE leading to protean clinical manifestations. In this case report, we present a 11-year-old Bangladeshi girl who presented with inflammatory pain in multiple joints, high-grade intermittent fever, headache, high blood pressure, and generalized tonic-clonic seizures. Detailed clinical examination with laboratory and imaging studies clinched the diagnosis of SLE and the involved organs include skin, mucosa, joint, haemopoietic system, kidney, and brain. In presenting this case and reviewing the literature, we emphasize the importance of very high index of suspicion for diagnosis of cSLE for early diagnosis, treatment and meticulous monitoring.

Keywords: Childhood onset systemic lupus erythematosus (cSLE), seizure, adult SLE (aSLE), glomerulonephritis, central nervous system.

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INTRODUCTION

SLE in children may present with multiple faces with variable course of the disease [1, 2]. Multiple organs/systems can be involved simultaneously or in a rapid sequence [1-3]. The incidence and prevalence of cSLE ranges from 0.36 - 2.5 /100,000 / year and 1.89 - 25.7 /100,000 respectively [4]. Presentation of cSLE among different ethnic groups has also variability [5, 6]. Reports are available showing cSLE presenting with more acute illness and having more frequent renal, hematological and neurological involvement within short course of time compared to adult SLE [7]. Reporting this case aims to emphasize the importance of very high index of suspicion for early diagnosis of cSLE, treatment and monitoring.

CASE SUMMARY

A 11-year-old Bangladeshi school girl presented with inflammatory pain in the multiple joints symmetrically involving small and large joints of upper and lower limbs for seven weeks, swelling of the face for three weeks, and headache for seven days. The swelling did not involve the other parts of the body, improved partially with diuretics, and was associated with reduction in volume and dark colorization of urine.

The headache was pulsatile, occasionally hemicranial, associated with nausea and vomiting; was not associated with increase in early morning, coughing, bending and straining; without any visual impairment, photophobia, redness of eye and lacrimation. During the course of illness, she had high-grade, intermittent fever, without shaking chills, persisted for initial three weeks. Exclusion of infection and non-response to empirical broad spectrum antibiotic led physicians to search for the presence of a multi-system connective tissue disorder. The fever subsided with initiation of glucocorticoid. She was admitted into hospital due to generalized tonic-clonic seizure nine times with tongue biting during the episodes of seizure three times. Infectious, metabolic and other causes of seizure were excluded. She had no significant past history and family history. She had not developed menarche still.

Examination revealed mild anaemia, high blood pressure (170/90 mmHg) and puffy face. As she was significantly improved on medication, clinical examination revealed normal other findings. Routine urine examination showed protein-(3+), red blood cells-20-30/high power field, WBC-plenty/high power field. Urinary total protein (UTP) was 1.7 g/24 hours. Full blood count showed features of pancytopenia. Anti-nuclear antibody (ANA) and anti-ds DNA were positive

in high-titer and complements were reduced. Serum creatinine, electrolytes and hepatic enzymes were within normal limit. Renal biopsy was advised but patient's parents did not agree to let their daughter to undergo biopsy procedure.

TREATMENT

The patient was dramatically improved after starting steroids. Initially soon after diagnosis, she was put on hydroxychloroquine (HCQ), oral prednisolone 0.5 mg/kg/day, and naproxen 250 mg 12 hourly. But 10 days after initiation of therapy she developed seizure attacks. She was hospitalized and received intravenous pulse methylprednisolone at 500mg daily for 3 days, followed oral prednisolone 1 mg/kg/day. She also received IV pulse cyclophosphamide 750 mg/square meter body surface area. Her blood pressure was controlled with captopril 50 mg and amlodipine 5 mg daily. Headache subsided with paracetamol. The patient was discharged home on oral steroids with follow-up appointments scheduled with rheumatology.

DISCUSSION

Systemic lupus erythematosus is the prototypic autoimmune disease characterized by heterogeneous, multisystem involvement and production of an array of autoantibodies leading to inflammatory tissue damage. The cSLE has an incidence ranging from 0.36 to 2.5 per 100,000 per year and a prevalence ranging from 1.89 to 25.7 per 100,000 [4]. The disease also shows a significant ethnic variability with higher incidence in African, Hispanic and Asian populations [5, 6]. Albeit uncommon, onset of pediatric SLE is described even in children younger than 2 years of age. The female to male ratio with pediatric SLE changes from 4:3 with disease onset during the first decade of life to 4:1 during the second decade to 9:1 in aSLE, and decreases to 5:1 in SLE commencing after the age of 50 year [14]. The data (summarized in Table-1) showed that common initial presentations of cSLE included constitutional symptoms, renal disease, musculoskeletal and cutaneous involvement. Less frequently involved at cSLE presentation were the neuropsychiatric, pulmonary and cardiac systems, with pericarditis reported in 3–24% of cases at presentation [15].

Table-1: Frequency of selected presenting clinical features of childhood-onset SLE

Balkaran <i>et al.</i> , [8] Spinosa <i>et al.</i> , [9] Abdwani <i>et al.</i> , [10] Hiraki <i>et al.</i> , [11] Agarwal <i>et al.</i> , [12] Gulay <i>et al.</i> , [13] Country Trinidad, Brazil, Oman, Canada, India and Philippines Sample size 33 47 50 256 70 78
Clinical features (%)
Malar rash 36 32 - 61 57 65
MSK#70 32 7661 66 41
Renal disease 64 38 64 45 77 63
Fever 76 34 62 39 94 -
Weight loss -* 26 52 29 30 -
Ulcers - 9 -33 - 54
Alopecia - 17 36 22 46 40
Pleuritis/Pleural effusion 9 17 26 12 3 14
Pericarditis/Peric-ardial effusion 24 4 22 12 3 15
Neuropsychiatric - 36 18 16 21 31

* denotes no data was presented for that clinical feature.

denotes musculoskeletal

Rashes occur frequently in cSLE, but only 30-50% manifest typical butterfly rash. Cutaneous lesions may take the form of photosensitive rash, recurrent urticarial, bullae, vasculitic nodules, or chronic ulceration. Discoid lupus erythematosus is unusual in cSLE [16]. Arthritis affects over 80% of children with SLE at some point. Usually, the arthritis involves the small joints of hand and feet with pain and swelling. Although the arthritis in cSLE is nondeforming, and responds well to anti-inflammatory medication significant arthritis at presentation is found in 40%-60% of children and adolescents [16].

The most common hematologic manifestation of cSLE is anemia. Usually the anemia is not coomb's positive hemolytic anemia with reticulocytosis; rather, it is a microcytic anemia of chronic disease. Leukopenia

and thrombocytopenia are also common but not invariably present [17].

Renal disease is evident in nearly two-thirds of patients with cSLE. Renal manifestations range from mild glomerulonephritis to sudden renal failure. Haematuria, proteinuria, and hypertension may be present in any combination. Renal biopsy should be performed if necessary to confirm the diagnosis, to investigate unexplained changes in renal function, and when the clinician is considering or monitoring the effects of aggressive therapy. Renal involvement is categorized according to criteria developed by World Health Organization. Mild clinical manifestation of renal involvement is usually well controlled with corticosteroid and diuretic agents. Persistent renal disease usually requires immunosuppressive therapy.

Chronic glomerular scarring is prevented by cyclophosphamide over the intermediate term. The systemic use of intravenous cyclophosphamide has been successful in children with diffuse proliferative glomerulonephritis and useful in membranous glomerulonephritis. More recently, mycophenolate mofetil has shown early promise, but its efficacy in routine practice is limited by the poor compliance. Monitoring of the patients with lupus nephritis is very essential. Adult series suggest that maintaining a creatinine clearance of 70 mL/min per 1.75 square meter is adequate [17].

Central nervous system involvement occurs in 20%-30% of children with SLE. Psychosis, personality change, seizure, chorea, transverse myelitis, peripheral neuropathy, and pseudotumor cerebri all may be the presenting manifestations. Delirium, hallucinations, seizures, and coma are most common objective neurologic signs in cSLE. The reported frequency of neuropsychiatric manifestations is lower in that in adults. Most often, central nervous system involvement occurs early in the natural course of cSLE. Frequently, it first become evident during or worsens immediately after the initiation of corticosteroid therapy. The explanation for this is uncertain, but these symptoms frequently resolve with pulse methylprednisolone therapy [17].

CONCLUSION

SLE is a multi-system autoimmune disorder. Besides significantly more active disease at the time of disease onset, there is also more active disease over time with pediatric SLE when compared to aSLE. Some children and adolescents with SLE are acutely ill at presentation. Multiple faces of aggressive lupus including fever, rash, joint pain, pancytopenia, haematuria, proteinuria, seizures, psychosis, sepsis etc may be present simultaneously or in a rapid sequence making a special challenge for the physicians to encounter the illness. High index of suspicion is necessary to diagnose the disease to initiate appropriate therapy.

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