Systemic lupus erythematosus and miliary tuberculosis in a prepubertal girl — a case report

S. Mahendra Raj, MBChB (Glasgow), MRCP (UK)
Lecturer

J. Hunt, MBBS (Monash), FRACP
Lecturer

Department of Medicine, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan.

Summary

A 13 year old girl presented with miliary tuberculosis and active systemic lupus erythematosus (S.L.E.). She responded to a combination of antituberculous drugs and systemic steroids. This case illustrates the fact that S.L.E. presenting in childhood may be rare but not unknown and exemplifies the need for vigilance in detecting life threatening infections in this group of patients.

Key words: Systemic lupus erythematosus, miliary tuberculosis, prepubertal girl.

Case Report

A 13 year old girl presented with a history of fever and generalised malaise of several week's duration. She was diagnosed as suffering from systemic lupus erythematosus (S.L.E.) two years before. On delving into the history it was apparent that the patient had a generalised skin rash since the age of seven years associated with relapsing and remitting facial erythema. She was admitted thrice over the preceding two years on account of episodic fever and recurrent pneumonia. On one occasion, vasculitic lesions were noted on her fingers, and she had bilateral non erosive arthritis of her ankles. She was thrombocytopaenic ($70,000/mm^3$) on her first admission. Since the time of diagnosis of S.L.E. she had intermittently been on oral prednisolone. She had not attained menarche.

Physical examination revealed a febrile, ill looking child who was clearly small for her age. She weighed 21kg and was 130cm tall. She had a generalised skin rash and alopaecia. There was an erythematous malar rash associated with areas of hyperpigmentation on the face. She had generalised icthyosis over her trunk and arms with localised areas of scarring. Tender matted lymph nodes were palpable in the neck, submandibular region, both axillae and groins. The liver and spleen were palpable 7cm and 5cm below the costal margin respectively.

Full blood count was unremarkable apart from a neutrophilia and total white cell count of 11.2 x 10^9/l. E.S.R. was 120mm/hr. Serum electrolytes were normal and liver function tests showed modest elevation of transaminases. Sputum specimens were repeatedly negative for acid fast bacilli on direct microscopy and culture.

The most significant finding was miliary mottling on the CXR and patchy opacities in the left lower zone. Review of serial films revealed that the left lower zone changes had been present on her previous admission. Bronchoscopy and broncheoalveolar lavage was undertaken to obtain bronchial washings, direct examination of which revealed acid fast bacilli on Ziehl-Neelson staining. Staining for *Pneumocystis carinii* was negative. Biopsy of a cervical lymph node revealed granulomatous caseation characteristic of tuberculous lymphadenitis.

The patient was started on antituberculous drugs consisting of Streptomycin, Rifampicin, Isoniazid and Pyrazinamide. She initially responded dramatically with defervescence of the fever and dimunition of systemic symptoms. However a week after the initial response the fever recurred and was associated with abnormal behaviour. The patient demonstrated aggressiveness and harrassed other patients in the ward. She was clinically judged to have had a recurrence of active S.L.E. and started on systemic steroids. This decision was vindicated by a prompt clinical response.

Results of serological autoantibody tests were available only after treatment was instituted. Antinuclear antibody was found to be positive in a titre of 1:320. Complement levels were low. Serum was negative for anti double-stranded DNA. C-Reactive protein was raised. Five months after starting treatment, the enlarged lymph nodes had resolved as had hepatosplenomegaly. It is notable that mental quotient as measured by the Wechsler memory scale was 64 indicating borderline mental retardation.

Discussion

The presence at some time or other of a rash, arthritis, acute psychosis, cutaneous vasculitis and thrombocytopaenia would generally be deemed sufficient to justify the diagnosis of S.L.E.[1] This case highlights the fact that S.L.E. presenting in prepubertal children maybe rare but by no means unknown.[2–4] The girl's poor physical and mental development could reasonably be attributable to S.L.E. The first clinical manifestations of the disease may precede diagnosis by years as indeed was the case in this patient who had the rash since the age of seven years.[3] This report exemplifies the need for vigilance in detecting life threatening infections when managing patients with SLE.

References

1. Tan EM, Cohen AS, Fries JF et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982; 25: 1271–7
2. Norris DG, Colon AR, Stickler GB. Systemic lupus erythematosus in children: The complex problem of diagnosis and treatment encountered in 101 such patients at the Mayo Clinic. Clin Pediatr 1977; 16: 774
3. Cassidy JT. Systemic lupus erythematosus. In: Cassidy JT ed. Textbook of Pediatric Rheumatology. Wiley Medical Publication, New York; 1982: 329–406.
4. Jonsson H, Niven O, Sturfelt G et al. Estimating the incidence of systemic lupus erythematosus in a defined population using multiple sources of retrieval. Br J Rheumatology 1990; 29: 185–8.