

posing to infection is a low granulocyte count(3). The present study shows a high incidence of bacterial infection in leukemia children even with normal granulocyte count. Empirical antimicrobial therapy is, therefore, recommended when these children are febrile even if the granulocyte count is normal.

Studies have demonstrated that total protective environment can reduce the number of infections in such children. However, the financial cost is prohibitive in a country like India. Psychologic stress and the emergence of resistant organisms are other disadvantages(4). Awareness of the problem in hospital personnel and parents and health education for possible preventive measures by hand washing and other hygienic measures may help. Prophylactic administration of antimicrobials is beneficial in this aspect. In a prospective study, ciprofloxacin was shown to be a better alternative to cotrimoxazole for preventing Gram negative infection in acute leukemias(5). However, -the administration of cotrimoxazole reduces

the incidence of *Pneumocystis carinii* infections.

In conclusion, infection especially by GNB is the major cause of mortality in childhood leukemias.

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## Myelofibrosis Secondary to SLE and its Reversal on Steroid Therapy

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Systematic lupus erythematosus (SLE) often causes hematological abnor-

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malities such as anemia, acquired immune hemolytic anemia, aplastic anemia, leucopenia and thrombocytopenia. Thrombocytopenia is usually attributed to peripheral destruction by platelet specific autoantibodies. This case reports our experience in myelofibrosis associated with SLE and its reversal after corticosteroid therapy.

### Case Report

A 12-year-old female known to have SLE since 1988, presented 3 years later with petechiae, fever and cough of 7 days duration. On examination, her vital parameters were stable. She had petechiae on the face and lower extremities. No organomegaly was detected.

Laboratory studies at presentation were as follows: Hb—9.6 g/dl, total WBC count— $4.7 \times 10^9/L$  with differential count of 14 polymorphonuclears, 70 lymphocytes, 12 monocytes and 4 atypical cells. ESR was 40 mm at the end of one hour. Platelet count was  $4 \times 10^9/L$  (Table I). PT and PTT were within normal limits. Direct Coomb's test was negative. Urinary analysis was normal. C3 was normal and. ANA (1:40) positive

though antids-DNA was negative. Liver and renal functions were normal. Bone marrow aspiration revealed normocellular marrow with decreased number of megakaryocytes. Other series were unremarkable. Bone marrow biopsy from the iliac crest revealed an extremely hypocellular marrow with residual patches of hematopoietic cells. Reticulin stain showed a marked increase in fine fibrosis which was diffuse grade III (Fig. 1).

Prednisolone was administered at a dose of 2 mg/kg/day. Petechiae disappeared within 2 weeks. Bone marrow aspiration and biopsy repeated after 4 weeks of treatment with steroids revealed a normocellular bone marrow with plenty of megakaryocytes. Myelofibrosis was fully reversed.

### Discussion

Myelofibrosis may be idiopathic or secondary to malignant and nonmalignant disorders(2). Growth factors released by platelets and megakaryocytes stimulate fibroblast proliferation and collagen synthesis. The factors implicated are platelet derived growth factor (PDGF); transforming growth factor-B

TABLE I—Hematological Parameters at Diagnosis and Follow-up

Parameter	At diagnosis	Day-8	Day-15	Day-24	Day-32
Hb (g/dl)	9.6	8.8	9.1	10.5	10.8
Retic (%)	1	9	2	1	1
TC ( $\times 10^6/L$ )	4.7	15.0	26.6	14.8	12.4
WBC (Polymorphs)	14	80	70	70	74
Platelet count ( $\times 10^9/L$ )	40	90	110	125	327

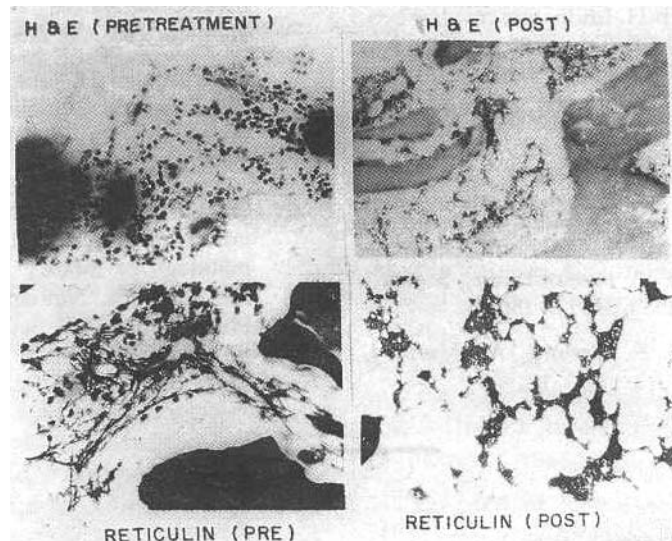


Fig. 1. Reversal of myelofibrosis in the bone marrow as demonstrated by reticulin stain before and after steroid therapy.

(TGF-B); and epidermal growth factor (EGF)(2). Besides, immunologic factors in the pathogenesis of myelofibrosis have also been suggested since patients with myelofibrosis have an increased incidence of autoantibodies and circulating immune complexes(1,3,4). An over-response of fibroblasts to autoimmune bone marrow damage or excess release of growth factors from platelets by binding immune complexes to Fc receptors of platelets is postulated. The reversal of myelofibrosis following immunosuppressive treatment supports the involvement of autoimmune bone marrow damage(5).

In SLE, platelet survival is shortened and there are increased numbers of bone marrow megakaryocytes. The platelet antibody is an IgG with molecular weight of 1.5-3.3 lakh daltons and binds complement. A qualitative defect

of platelet function also may be present. Patients with SLE may have a serum inhibitor of platelet aggregation(6).

Corticosteroid therapy in our case was effective in reversing both the hematological manifestations as well as the myelofibrosis. This strongly suggests that myelofibrosis in this patient was caused by SLE. Thrombocytopenia could have been due to a combined effect of autoimmune process and myelofibrosis.

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## Hypophosphatemic Rickets with Hypercalciuria

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A new syndrome, hereditary hypophosphatemic rickets with hypercalciuria (HRH), was recently described in closely related members of a Bedouin tribe in Israel(1,2). This condition is characterized by renal phosphate leak resulting in hypophosphatemia with an

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appropriate elevation of blood levels of 1,25 dihydroxyvitamin D, increased intestinal calcium absorption and hypercalciuria. HRH is a distinct condition and must be differentiated from the patients of classic hypophosphatemic rickets. The rarity of this condition prompts us to report the clinical and laboratory features and response to therapy in two such patients.

### Case Reports

*Case 1: A 6-year-old girl of unrelated healthy parents was referred for evaluation of long standing rickets. She was well till the age of one and a half years when bowing of legs and difficulty in walking were noticed. The bony deformities chiefly involved the lower limbs and progressively increased. At the age of 5 years she started complaining of bone pain, mainly at the pelvic girdle and legs, and developed a waddling gait. On examination the height was 102 cm (<5th percentile) and weight 17 kg*