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Reply

Firstly, we would be very skeptical in making a diagnosis of SLE without fulfillment of the 1982 revised criteria for SLE. Our report was focused on an unusual complication of SLE in a known patient diagnosed to have SLE 3 years prior to this hematological manifestation. Obviously when the diagnosis of SLE was made in 1988, this girl clinically fulfilled the required criteria. She had skin rash, arthritis, CNS and renal manifestations at that time which remitted on steroid therapy. Then she was lost to follow for about 3 years and presented again with the hematological manifestations as described in our report (she came from a remote place in Gujarat). The mother reported that the child was apparently well during this period. Thus, in this child a diagnosis of SLE was never in doubt; whether SLE was active at the time of detection of myelofibrosis could be debated.

SLE is known to have spontaneous exacerbations and remissions. Also children with milder disease may not report for regular follow up. It seems that our patient had myelofibrosis as a hematological disorder representing exacerbation of SLE. She had anti-DNA antibody in abnormal titers. Anti-Sm and anti-ds-DNA antibodies may be negative with normal levels of serum complement, even with active systemic SLE in the absence of nephritis (as with our patient). Regarding the gap between this child's hematological disorder and the diagnosis of SLE, there have been a number of reports corroborating this fact(1-7). It is interesting that most of these previous patients (including the present case) lacked many of the classical, clinical SLE symptoms at the time of the diagnosis of myelofibrosis(4).

Secondary, as regards presence of megakaryocytes (MK) in the bone marrow aspirate and biopsy, acute or malignant myelofibrosis would be associated with increased MK numbers whereas in secondary myelofibrosis, findings are generally variable(8). If SLE is associated with immune peripheral destruction of platelets, then definitely the bone marrow MK are increased in number. In our patient, there were no demonstrable anti-platelet antibodies or any other evidence for immune destruction of platelets. Additionally, the bone marrow biopsy was hypoplastic in this child with decreased numbers of MK.

Finally, regarding the suggestion of an EBV infection in this child, there was no clinical or laboratory evidence for EBV infection. Children with SLE are prone to infections, but EBV infection would rarely present with a petechial rash or pancytopenia at initial diagnosis as

happened in this child.

In conclusion, we emphasize that there was no doubt about the diagnosis of SLE in this child. The recognized features of active SLE may have been lacking at the time when she developed myelofibrosis. But the reversal of myelofibrosis within 4 weeks or oral steroid therapy clinched the issue and tilted the scales in favor of our proposition. This case is an unusual presentation of myelofibrosis secondary to SLE !.

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