

Myelofibrosis Secondary to SLE and its Reversal on Steroid Therapy

With reference to the aforementioned case report(1), We would like to point out some aspects which go against the diagnosis of SLE, as the patient doesn't satisfy the 1982 revised criteria for the classification of SLE, where a minimum of 4 out of 11 criteria are taken into account for the diagnosis of SLE(2). Kidney involvement can be an isolated feature.

In this case, a diagnosis of SLE was made in 1988 and the patient landed up with myelofibrosis after 3 years, with 8 days history of cough, fever and petechiae. In this context, the following pertinent aspects need elaboration: (i) The minimum diagnostic criteria are lacking; (ii) Status of SLE is not described during the 3 years interval from 1988; (iii) It is not mentioned whether the patient was on maintenance dose of steroid or any other drugs. Perhaps, the patient was not on treatment, as steroid itself reversed myelofibrosis, or is it a fact that myelofibrosis developed suddenly as an exacerbation of SLE? In that event anti ds-DNA should have been positive, which is virtually diagnostic of active SLE and exacerbation of SLE(3); (iv) ANA, which is positive, can be observed in many other conditions(2); (v) ESR is markedly raised in SLE, but in this case it was just 40 mm; (vi) Kidney involvement is absent; and (vii) It is not clear whether myelofibrosis was secondary to some unrecognized agent or just idiopathic. In such instances, estimation of urinary excretion of hydroxyproline, this is increased in secondary myelofibrosis, would have helped(4).

Daly and Scotts reported a case of pancytopenia due to myelofibrosis secondary to active SLE in 1983(5). In this case, the bone marrow revealed marked fibrosis with plentiful of megakaryocytes. Other related literature also confirms this finding(4,6) and even in SLE, there are increased number of megakaryocytes in the bone marrow(7). However, the authors of the case report under discussion(1), reported a decreased number of megakaryocytes in the bone marrow, which needs explanation.

In the present case, features like mild anemia, granulocytopenia, thrombocytopenia, lymphocytosis, presence of ANA and atypical lymphocytes and reversal of pathology with steroid are also compatible with a diagnosis of EBV infection (though counts of atypical lymphocytes are low).

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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