AUTOIMMUNE MYELOFIBROSIS ASSOCIATED WITH LUPUS: UNUSUAL OR UNDETECTED?

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Systemic lupus erythematosus (SLE) is a systemic autoimmune disease more commonly affecting young females. Hematological manifestations are one of the important clinical features of patients with SLE. Hemolytic anemia, leucopenia (predominantly lymphopenia), thrombocytopenia, and less commonly pancytopenia may be found in up to four-fifth of patients with SLE and form part of the classification criteria commonly used for lupus [1, 2]. Autoimmune myelofibrosis (AIMF) associated with lupus is one of the less common scenarios reported to cause cytopenias in SLE. In this issue of the Journal, Cansu *et al.* propose that AIMF might not be as rare as it has been described in the literature, and might actually be underrecognized [3].

AIMF causing cytopenias (generally pancytopenia) is one of the severe manifestations associated with lupus. In such instances, severe cytopenias involving all three lineages can be life-threatening. AIMF might require multiple transfusions of blood product units until (and if) there is recovery of the process of myelofibrosis following immunosuppressive therapy. Refractoriness to immunosuppressive therapy might require bone marrow transplantation as a lifesaving measure. The severity of AIMF is precisely the reason why it remains unlikely that

AIMF is underrecognized in lupus. Patients with pancytopenia in the context of lupus would generally undergo bone marrow examination. In such instances, a hyperactive bone marrow would indicate peripheral destruction of erythrocytes, leucocytes, and thrombocytes. On the other hand, a hypocellular marrow would indicate marrow suppression, whether related to antibodies to stem cell precursors, gelatinous transformation of bone marrow, or myelofibrosis resulting in myelophthytic anemia [1, 4-6]. This is precisely the reason why the hypothesis that AIMF is underreported (rather than uncommon) might not be valid.

It is still possible that AIMF might be underreported in the literature owing to its rarity and the difficulty in publishing isolated case reports in the present publishing scenario. Therefore, the hypothesis proposed by Cansu *et al.* still requires to be tested in real-life situations to assess its validity [3]. A first step towards testing this hypothesis could involve sending questionnaires to consultants leading lupus clinics across a geographic region or regions. Such questionnaires could assess what proportion of patients from these clinics have actually been suspected to have AIMF and how many actually underwent a bone marrow examination to look for

myelofibrosis. Should the frequency of myelofibrosis in lupus detected by such means far exceed that in the reported literature, then such findings should be prospectively verified in longitudinal cohorts of patients with lupus. As the authors have rightly proposed [3], stringent criteria delineating the need for bone marrow examination in patients with lupus might help detect rarer hematological manifestations such as AIMF more frequently. However, as pointed out previously, the overall prevalence of AIMF in lupus is still likely to be low

due to the fact that severe pancytopenia *per se* is rare in lupus.

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Durga Prasanna Misra declares that he has no conflict of interest, including no relationship with pharmaceutical companies.

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**ЛУПУСПЕН БАЙЛАНЫСТЫ АУТОИММУНДЫ МИЕЛОФИБРОЗ: СИРЕК НЕМЕСЕ
АНЫҚТАЛМАЙТЫН МАНИФЕСТАЦИЯ?**

Түйінді сөздер: аутоиммунды миелофиброз, диагностика, гипотезалар, жүйелі қызыл жегі.

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