

Systemic Lupus Erythematosus in a Patient with Essential Thrombocytemia

Esansiyel Trombositemi'li bir hastada Sistemik Lupus Eritematozus

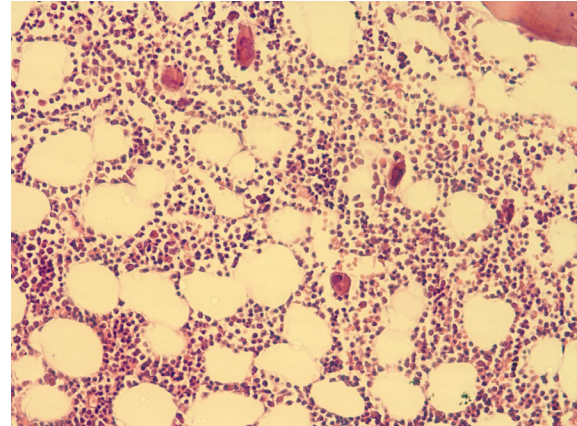
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Dear Editor;

It is possible to see various hematological abnormalities in systemic lupus erythematosus (SLE). These abnormalities include anemia, thrombocytopenia, and leucopenia (1). Thrombocytosis is a condition in which there is a number of platelets over 450000 in the blood (2). This can stem from the reactively stimulation of platelet production in various diseases (iron deficiency anemia, malignances, infections, postsplenectomy or asplenic conditions), familial mutations, essential thrombocytemia, and other myeloproliferative disorders (3). Thrombocytosis due to autosplenectomy is an unusual hematologic finding in SLE. Essential thrombocytemia (ET), one of the reasons for thrombocytosis, is one of the chronic myeloproliferative disorders which JAK 2 gene mutation can be detected, and it is characterized by thrombocytosis and abnormal proliferation of megakaryocytes. Here, we have presented a patient who was observed with the coexistence of ET and SLE.

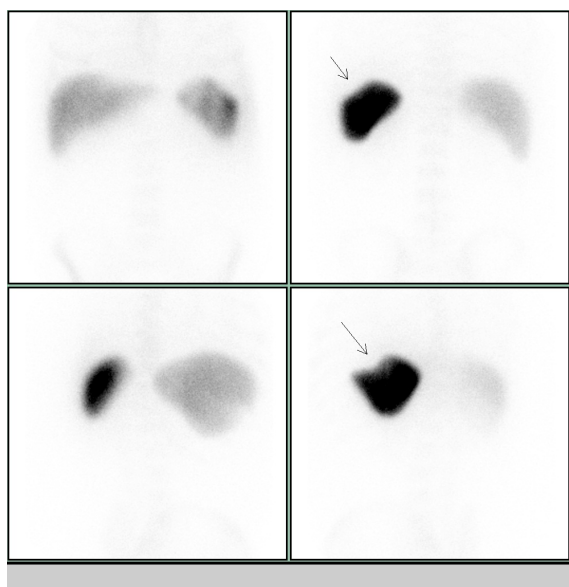
69 years old male patient complaining of recurrent epistaxis was admitted in May 2007. Physical examination of the patient was not remarkable. No pathology explaining the epistaxis was detected in the ear, nose, and throat examination. Urinalysis, liver and kidney function tests were normal. The patient's PT and aPTT values were normal as well. The number of platelet was found to be $1.200 \times 10^3/\mu\text{L}$. Peripheral smear of the patient was not remarkable except for thrombocytosis. Possible causes of reactive thrombocytosis were excluded. While hyperplasia was detected in the megakaryocytic series in bone marrow biopsy, reticulin fibrosis, erythroid hyperplasia and neutrophil granulopoiesis were not observed (Figure 1).



Jak2 gene mutation was positive, ET was accepted and hydroxyurea treatment was started. The patient readmitted in June 2009 with pain in hand joints, swelling, temperature rise, photosensitivity, and malar rash. The erythrocyte sedimentation rate was 100 mm/h, leukocyte count was $5600/\text{mm}^3$, lymphocytes $800/\text{mm}^3$, and platelets $450 \times 10^3/\mu\text{L}$. ANA was found to be 1/100 nucleolar, anti-dsDNA was 78.5 IU/ml, and accordingly, the patient was diagnosed as SLE (4,5). It was thought that the autosplenectomy as the factor for SLE associated with secondary antiphospholipid syndrome in thrombocytosis etiology. Lupus anticoagulant, antikardiyolin IgM and IgG were negative. While the performed peripheral smear to determine autosplenectomy was normal, only a little splenic infarct was observed in the captured Tc-99m heat denaturated red blood cell (RBC) scintigraphy (Figure-2).

Essential thrombocytemia is a disease which is characterized by abnormal increase in bone marrow megakaryocytic series. Peripheral blood is normal except for thrombocytosis. The bone marrow biopsy is necessary for the diagnosis of ET although some investigators claim

that it is insignificant (6). The bone marrow biopsy in this case supports the diagnosis of ET.



Regarding SLE, just as thrombocytopenia is seen frequently in platelet abnormalities, thrombocytosis may rarely occur. The presence of persistent thrombocytosis especially in the SLE patients should remind autosplenectomy due to antiphospholipid antibody syndrome (7). In this case, evaluations indicated that there were neither any findings of antiphospholipid syndrome, nor peripheral smear and scintigraphic findings of autosplenectomy. The etiology of thrombocytosis remains unknown in the majority of SLE cases with persistent thrombocytosis. In related case reports, only three were identified to cause autosplenectomy due to antiphospholipid antibody syndrome (8). From this perspective, our case is the first in the literature to show the coexistence of SLE and ET. When other cases in the literature are taken into account, the possibility of ET should also be kept in mind in the presence of persistent thrombocytosis in SLE patients.

References

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