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Systemic Lupus Erythematosus with Pancytopenia, a challenging case.

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Introduction: Hematologic manifestation is a challenging presentation in systemic lupus erythematosus (SLE). It has been a well-known systemic autoimmune disease with hematologic abnormalities such as anemia, leukopenia, and thrombocytopenia. Myelodysplastic syndrome (MDS) is defined as a clonal expansion of Bone marrow derived pluripotent stem cells. Few cases of co-occurrence of two diseases have been reported. Here is a patient with SLE with hematologic manifestations who are eventually diagnosed with the MDS.

Patient History: A 46-year-old woman known case of SLE, with a history of chronic renal failure from 10 years ago was referred by a hematologist due to pancytopenia. Some time ago, she was admitted to the hematology service due to pancytopenia and at that time she was taking azathioprine and allopurinol simultaneously.

The patient was a middle-aged woman with a Cushing's appearance and had no specific clinical complaints.

CBC: WBC: 3500, Hgb: 7.3, Plt: 76000, ESR: 76, Serum Cr:2.15 mg/dl, uric acid: 6.8 mg/dl.

ANA:10.1(1.1), dsDNA: neg, complement levels: not decreased. Antiphospholipid Ab: neg and 630 mg protein in 24 h urine.

Treatment with prednisolone, hydroxychloroquine and cellcept were started. One month later, the results of lab data included:

CBC: WBC: 12000, Hgb: 8.6mg/dl, Plt: 150000, ESR: 17, Serum Cr:1.5 mg/dl.

Two months later, she experienced rectal bleeding due to hemorrhoid and Hgb decreased to 5.5 mg/dl. Blood transfusion was prescribed, but anemia wasn't corrected and then Plt was decreased too. Dose of cellcept was decreased and high dose of prednisolone was prescribed, but it wasn't effective. Finally bone marrow aspiration and biopsy were done and was compatible with refractory cytopenia with multilineage dysplasia (MDS). Treatment by erythropoietin was added to treatment.

discussion: There are some case reports about association of MDS and rheumatic disorders. In this patient, concomitant use of azathioprine and allopurinol, renal failure, rectal bleeding, and finally MDS were different causes of cytopenia.

Mobini et al have evaluated 80 patients with MDS and 9 (11.3%) patients were diagnosed as having rheumatic disorders, one of them was SLE. In younger patients, refractory cytopenia and refractory cytopenia with multilineage dysplasia were more prevalent [1].

Jung et al have reported four patients with both MDS and SLE, they were all women from 15 to 50 years old, with anemia or pancytopenia and bone marrow findings were disclosed as MDS[2].

Due to the variety of causes of hematologic involvement in SLE, less common causes such as MDS should be considered in resistant or unusual cases.

Key word: systemic lupus erythematosus, pancytopenia, myelodysplastic syndrome.

References

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