[症例報告]Severe Anemia with Autoimmune Thrombocytopenia : a Case Report of Possible Sytemic Lupus Erythematosus

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Severe Anemia with Autoimmune Thrombocytopenia; a Case Report of Possible Sytemic Lupus Erythematosus

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Abstract

We report a case of severe thrombocytopenia with anemia. The thrombocytopenia was associated with a decrease in megakaryocytes in bone marrow and immunological abnormalities suggesting systemic lupus erythematosus, and was improved by steroid therapy. Autoimmune thrombocytopenia as possible prodrome for systemic lupus erythematosus was discussed.

Key words: thrombocytopenia, systemic lupus erythematosus, anemia, petechia, steroid therapy

Introduction

Systemic lupus erythematosus(SLE) is known to present several hematologic abnormalities[1], and autoimmune thrombocytopenia can be a prodrome for this disease[2].

We report here a woman showing severe thrombocytopenia associated with immunolgical disorders suggesting latent SLE.

Case Report

A 31-year-old woman was admitted to the Ryukyu University Hospital in December,1986 because of exertional dyspnea and palpitation. She was healthy until the age of 26, when a slight anemia was found during her pregnancy for the first time. However, she received no treatment for the anemia. Two years ago she had a general fatigue and was transfused of blood for anemia in another hospital. One year prior to admission, she began to cause massive menstrual bleeding and frequent hemorrhages from the gum. She has experienced palpitation and shortness of breath on exertion since two months earlier, and was admitted to our hospital. There were no past and family histories related to this condition.

Physical examination on admission revealed a well-nourished Japanese woman with moderate

stature of height 149 cm and weight 42.5 kg. The body temperature was 36.7°C. The arterial blood pressure was 120/70 mmHg, and the pulse rate was 84/min. The skin was pale, the palpebral conjunctiva was anemic, and the bulbar conjunctiva was not icteric. There was no skin lesion such as butterfly rash, discoid lupus, alopecia, or photosensitivity. The salivary and lacrimal secretion were not impaired. No lymphadenopathy was found. The jugular venous pressure was not increased. A venous hum was heard on the lateral neck. The thyroid goiter was not felt. The lungs were clear; rale, rhonchi, or pleural friction rub was not heard. A grade 3 systolic murmur was heard along the lower left sternal border and the cardiac dullness was enlarged. The abdomen was normal; the liver and spleen were not felt. There was no peripheral edema, clubbing, nor cyanosis. No tenderness nor limitation of motion was observed in all joints. Rectal examination was negative.

The hemoglobin at this time was 3.4 g/dℓ, hematocrit was 13.4%, red blood cell count was $195 \times 10^4/mm^3$, the white blood cell count was $3,000/mm^3$, with 71% neutrophils, 25% lymphocytes, 1%monocyes, and 2% basophils and platelet count was $12,000/mm^3$. The prothrombin time was 15.8 seconds with a control of 14.4 seconds; the partial thromboplastin time was 46.1 seconds with a control of 40.9 seconds. The fibrin-fibrinogen degradation product(FDP) was negative, and fibrinogen was 227 mg/dℓ. The total iron binding capacity was 462 μg/dℓ, serum Fe was 13 μg/dℓ, and ferritin was 9.5 ng/dℓ. Bone marrow showed a decrease in megakaryocyte count and a various size of platelets without dysplastic cell. On the serological examination, LE test was positive, RA test was positive, Anti-DNA antibody was a titer of 1:1280, anti-ENA antibody was positive, SS-A antibody was a titer of 1:16, anti-platelet antibody was positive, anti-microsome antibody was a titer of 1:102,400, and anti-thyroglobulin antibody was a titer of 1:6,400. However, anti-SM antibody, anti-RNP antibody, SS-B antibody, scl-70 antibody, and anti double-strand DNA antibody were negative. The direct Coombs' test was negative, IgG was 3,000 mg/dℓ, IgA was 435 mg/dℓ, IgM was 178 mg/dℓ, and the hypocomplementemia was present; C3 was 100 mg/dℓ, C4 was 100 mg/dℓ, and CH50 was 26 U/mℓ. There were no remarkable abnormalities in urinalysis, renal function tests, and liver function tests.

Chest roentogenogram demonstrated cardiomegaly with 60% of cardiothracic ratio and the lung field was clear. An electrocardiogram demonstrated a left ventricular overload with a normal rhythm at a rate of 62. An echocardiogram revealed a dilataion of the left ventricle.

During the period of hospitalization, she was treated with ferrous sulfate and her hemoglobin rose to 10 g/dℓ within one month. The symptoms disappeared and the cardiomegaly was improved on the chest X- ray film. Despite the correction of the anemia, she began to have petechiae on the extremities and back. Rumpel-Leede test was positive with bleeding time of 7 min, and platelet count $6,000/mm^3$, while her prothrombin time and the partial thromboplastin time were not prolonged. FDP was negative, and the fibrinogen was 301 mg/dℓ. No massive bleeding was apparent. The patient was treated with a first daily dose of 60 mg of prednisolone, and there was a striking clinical improvement in hematologic and immunologic findings; the platelet count was increased to $100,000/mm^3$, the titer of anti-DNA antibody was reduced, the serum level of IgG was decreased to 1,480 mg/dℓ, and the hypocomplementemia was improved. Subsequently the steroid was tapered slowly to the maintenance dose(Fig.1).

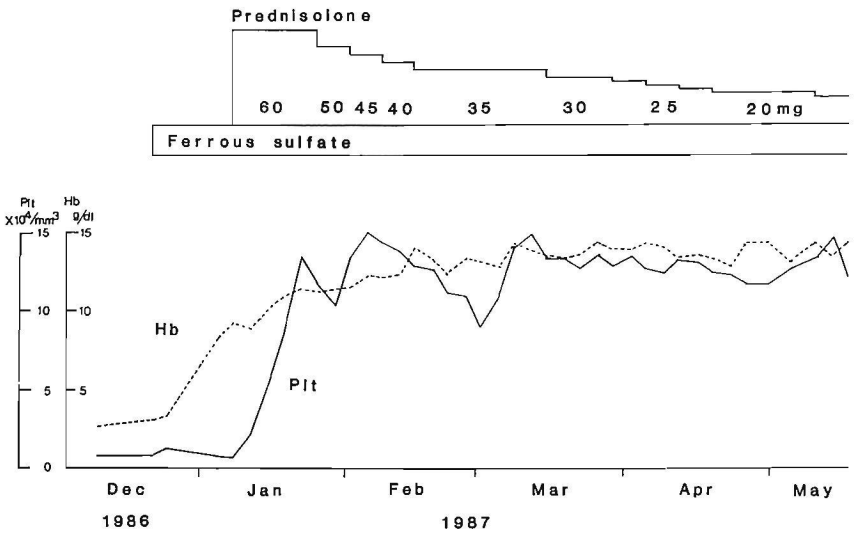

Fig.1 Clinical course of the patient. Anemia was improved by the administration of ferrous sulfate. Addition of prednisolone markedly increased the platelet count. Hb:hemoglobin concentration, Plt:platelet count.

Comments

SLE is a connective tissue disease which is characterized by many organ involvements and marked and varied immunologic abnormalities, including a variety of autoantibodies participating in tissue injury[3] . One or more hematologic abnormalities are present in nearly all patients with active SLE.[4] Although anemia is the most common hematologic abnormality[1] , thrombocytopenia occurs in 6.9 to 14% of patients with SLE[4~6] .

In this case, the marked thrombocytopenia was a initial clinical manifestation. Her severe anemia should be caused by blood loss secondary to the thrombocytopenia, because the only administration of ferrous sulfate markedly improved the anemia. The thrombocytopenia was still present after the improvement of the anemia, therefore, it should be considered to be one of the primary abnormalities in this case.

Platelet counts can be decreased by many causes such as drugs, inflammatory diseases, malignant neoplasms, primary hematologic disorders, or autoimmune mechanisms. However, she had no evidence of the above mentioned disorders except for the immunological abnormalities. Since anti-platelet antibody was detected, we should regard idiopathic thrombocytopenic purpura(ITP) as a differential diagnosis of the thrombocytopenia. ITP may be characterized by increased number of megakaryocytes in the bone marrow, sole thrombocytopenia, and absence of any other disease known to give rise to thrombocytopenia[5] [7] .

This patient had leukocytopenia and decreased megakaryocyte counts in the marrow. The many immunological abnormalities found in this case may suggest that patient suffered from connective tissue disease, especially SLE, though the manifestaions in the patient could not conform to The 1982 Revised Criteria for Classificfation of Systemic Lupus Erythematosus. The immune thrombocytopenia

can be a prodrome for SLE[2], although it was rarely reported to the initial manifestaion of SLE[4]. Furthermore, the bone marrow finding and the hypocomplementemia were compatible with the findings in SLE. The other connective tissue disease, such as Felty syndrome or Sjogen syndrome, can cause a decrease in platelet count, but such a syndrome might not be considered because of the absence of splenomegaly, sicca syndrome, or objective arthritis.

In summary, our patient was a relatively rare case with possible SLE initially developed with severe thrombocytopenia and anemia.

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