

Megakaryocytic Aplasia

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Abstract

Acquired Megakaryocytic aplasia is a rare disorder defined by severe thrombocytopenia with no other haematological and absent or severely marrow megakaryocytes.

Key Words

Megakaryocytic Aplasia, Mycophenolate mofetil.

Introduction

Acquired megakaryocytic aplasia is a rare disorder defined by severe thrombocytopenia with no other haematological abnormalities and absent, or severely decreased marrow megakaryocytes (1). Until recently, less than 20 cases of acquired megakaryocytic aplasia have been reported and the disease etiology remains unclear (2).

Case Report

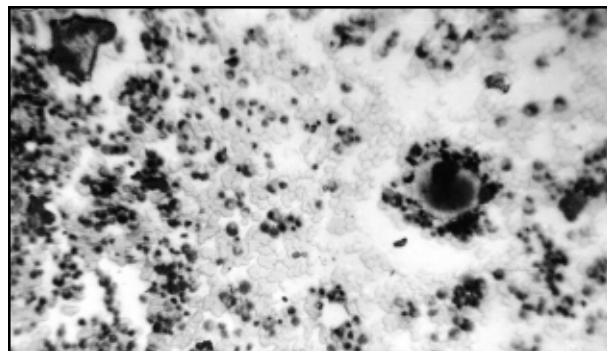
A 23 year unmarried female was admitted with history of menorrhagia, continuous bleeding and passage of clots for the last 15 days. One month back, there was an episode of epistaxis for which she was hospitalized and received blood transfusions. There was no history of trauma, drug intake, fever or rash and no family history of bleeding diathesis.

On examination, the weight was 42 kg. The patient was conscious and afebrile. Marked pallor was present. Pulse rate was 96 per minute, blood pressure 100/60 mm of Hg. There was no significant lymphadenopathy. Cardiovascular, respiratory and abdomen examination were normal.

Investigations revealed hemoglobin of 7.4 g/dl with total leukocyte count 6200 per mm³ with 64% neutrophils, 32% lymphocytes, 2% monocytes, 2% eosinophils. BT - 2'.00", CT - 4'.50", blood group A+ve

and platelet counts 20000/mL. Peripheral blood film showed normocytic hypochromic red blood cells with moderate degree of anisocytosis and poikilocytosis. Bone marrow examination showed gross reduction in the number of megakaryocytes. ESR was 25 mm in first hour by Westergren Method. Biochemistry revealed fasting blood sugar 100 mg/dl, urea 31 mg/dl, creatinine 0.6 mg/dl, total bilirubin 1 mg/dl. Alkaline phosphatase 150, SGOT - 53, SGPT - 42. Chest radiograph and USG abdomen was also normal. HBsAg, anti-HCV, Abs and HIV test were negative. ANA levels in normal range.

A final diagnosis of acquired megakaryocytic aplasia was made. Patient was given blood transfusion, platelet transfusion and dexamethasone. Patient improved and was discharged from the hospital after 2 weeks. Patient is on regular follow-up and doing well.



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Discussion

Acquired amegakaryocytic thrombocytopenia (AAT) is a rare disease characterized by severe thrombocytopenia due to selective reduction or absence of megakaryocytes in the bone marrow (3, 4). It may be a primary disorder or may be seen in aplastic anemia (AA), preleukemia, systemic lupus erythematosus (SLE) and nutritional B12 deficiency (5, 6, 7). More commonly, patients with acquired amegakaryocytic thrombocytopenia have additional hematologic abnormalities such as macrocytosis or dyserythropoiesis, abnormalities which may predict progression to aplastic anemia or myelodysplasia.

Acquired megakaryocytic aplasia is a disease of hematopoietic stem cells manifesting as thrombocytopenia which subsequently may progress into aplastic anemia or myelodysplasia. The patient may be treated with corticosteroids, lithium carbonate, androgens, vincristine, immunosuppressive drugs, platelet transfusions and plasma substitution.

Some study trials evaluated immunosuppressive treatment of AAT with antilymphocyte globulin (ALG) or antithymocyte globulin (ATG), combined with cyclosporine showed effectiveness in patients (8). Danazol has been also tried in some patients. Recently, mycophenolate mofetil was used to treat a single case of acquired megakaryocytic aplasia (9).

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