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Acquired Pure Megakaryocytic Aplasia: Report of a Single Case Treated With Mycophenolate Mofetil

Deepti Bulchandani,[1]* Jagdish Nachnani,[1] Robert Belt,[2] and Stuart Hinton[2]

[1] Department of Internal Medicine, University of Missouri-Kansas City, Kansas City, Missouri
[2] Department of Hematology/Oncology, Kansas City Cancer Center, University of Missouri-Kansas City, Kansas City, Missouri

Acquired pure megakaryocytic aplasia (APMA) is a rare disorder in the field of hematological diseases. We report the case of a patient with APMA treated with mycophenolate mofetil and review the literature associated with this. Until today, very few cases of APMA have been reported and the disease etiology still seems to be unclear. There have been no reported cases of treatment of APMA treated with mycophenolate mofetil. Am. J. Hematol. 82:650–651, 2007. © 2007 Wiley-Liss, Inc.

Key words: acquired pure megakaryocytic aplasia (APMA); mycophenolate

INTRODUCTION

Acquired pure megakaryocytic aplasia (APMA) is a rare disorder characterized by severe thrombocytopenia with no other hematological abnormalities [1,2]. The exact etiology and pathogenesis behind this disease is still uncertain [3]. Intensive immunosupression using cyclosporine combined with antithymocyte globulin has been shown to be effective in patients with aplastic anemia who do not undergo bone marrow transplantation [4,5] and a similar regimen has been described in treatment of two patients with APMA [2]. We report the first case of APMA treated with mycophenolate mofetil.

CASE REPORT

We present the case of a 70-year-old Caucasian lady who has presented with a several month history of increased bruising. This occurred in the setting of her taking aspirin and Coumadin for a history of atrial fibrillation. She has never had any bleeding or clotting issues. Her family history was basically noncontributory.

Platelet count done on October 2005 was 11,000 and 4 days later was 10,000. Her other cell lines were normal and her previous platelet counts had been normal so it was assumed this was idiopathic thrombocytopenia purpura and she was started on prednisone 1 mg/kg.

Her platelet count did not respond promptly. Bone marrow aspirate biopsy done on November 1, 2005 showed no major changes in the white and red cell line, but she did have basically absent to markedly decreased megakaryocytes. Flow cytometry was unrevealing and cytogenetics was normal. Parvovirus titers were consistent with old infection but not active. Her antiplatelet antibody was also negative. Her B12 and folate levels were normal and her TSH was normal. She had no obvious reason for this. She was on a thiazide diuretic which was stopped and she had taken penicillin some time before for a urinary tract infection. She was also on Valsartan, digoxin, atenolol, potassium chloride, thyroxine supplement, fluoxetine, calcium, and vitamin supplement. She denied the use of any herbal medicines or alcohol. We followed her for over 6 weeks though and did not see any evidence of improvement. Her steroids were tapered off after the bone marrow biopsy. The patient was followed over the next month with platelet counts ranging between 6000 and 14000 range and no significant change in the white blood cell count or hemoglobin. A second bone

*Correspondence to: Deepti Bulchandani, 6911 W 52nd Place Apt No. 3B Mission, Kansas, KS. E-mail: dbulchandani@yahoo.com

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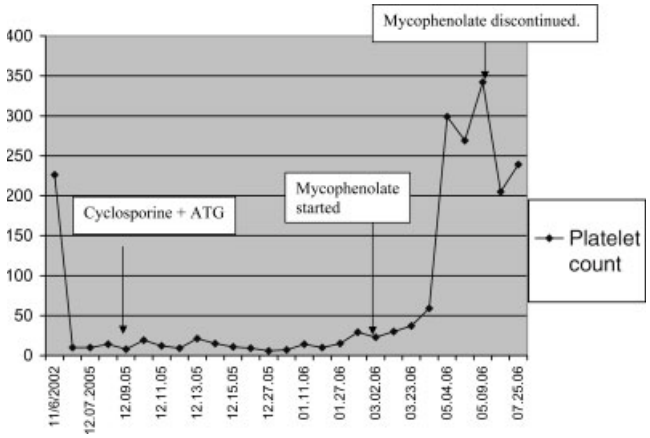

Fig. 1. Graph showing the sequential changes in platelet counts.

marrow aspirate and biopsy done on December 7, 2005 showed findings similar to the first bone marrow biopsy. We elected to proceed with immunosuppressive therapy, and she received a combination of rabbit ATG at 3.5 mg/kg/day for a total of 8 days in December from December 8th to December 15th. She was also started on cyclosporine on December 8, 2005, a total of 500 mg twice daily and prednisone. She subsequently was admitted with confusion which was thought to be due to cyclosporine and prednisone neurotoxicity. Her cyclosporine and prednisone was initially discontinued and her mental status changes improved. She was rechallenged with cyclosporine in mid-January and she again started developing mental status changes and pulmonary infiltrates which responded to steroids. Her platelets were still in the 20 thousands. At this time we decided to start her on mycophenolate mofetil on March 2, 2006 at which time her platelet count was 23,000.

Her platelet counts subsequently improved (Fig. 1). She was admitted to our hospital in May 2006 for an unrelated partial small bowel obstruction and an episode of zoster limited to her neck during which time her platelet count had normalized. We discontinued CellCept because of zoster and her repeat platelet count has remained normal.

Review of Literature

We did a review of literature looking for case reports of APMA. The databases searched were MEDLINE and OVID. The Search terms used were: Acquired pure megakaryocytic aplasia, APMA, Megakaryocytic aplasia.

DISCUSSION

While isolated thrombocytopenia after toxic exposure or with an immunogenic cause is relatively common and immunosuppressive treatment is effective, acquired pure megakaryocytic thrombocytopenia remains a rare disease in the field of hematological disorders [1]. The clinical course of this rare disease seems to be variable [6]. In some patients it progresses rapidly to aplastic anemia [7].

There has been case reports of APMA treated with ATG and cyclosporine in a few patients [2,7,8], though there have been anecdotal reports of treatment failures with this regimen. Mycophenolate mofetil has never been described in the treatment of APMA. APMA is a disease with high mortality and our case report demonstrates that mycophenolate may be considered in patients of APMA who are intolerant of cyclosporine.

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