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Allogeneic Bone Marrow Transplantation from Haplomatched Sibling Donor in a Pediatric Patient with Sickle Cell Disease with Multiple Painful Crises: An Experience

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Editorial

Bone marrow transplantation is the only cure for sickle cell disease but this curative therapy unfortunately lead to complication and several times death. Bone marrow transplantation indicated only for sickle cell patients with severe sickle cell disease who have complications including strokes, acute chest syndrome, recurrent pain crisis and exchange transfusions, nephropathy retinopathy, osteonecrosis of multiple joints and priapism. All sickle cell patients are not eligible for bone marrow transplantation due to associated toxicity [1]. Transplant is performed when benefit outweighs the risk. There has been much development and improvement in the bone marrow transplant since the first transplant done in 1984. This technique includes using chemotherapy for conditioning to remove the recipients cells and replacing it with donor cells free of sickling. Main problem limiting the transplant to all patients include getting a matched donor, risk associated with the procedure; including graft *vs.* host disease and cost. Hematopoietic stem cell transplant is safest when a matched donor is available. Even with this, there is still risk of graft rejection and chronic graft verses host disease. The unrelated cord blood stem cell transplant and haploidentical stem cell have been found to be less successful due to an increase in graft *vs.* host disease. Bone marrow transplantation has helped in improving the quality of life of the patient by eliminating the repeated hospital admission [2].

We report a diagnosed case of sickle cell anemia, 10 years old male child with repeated episodes of pain crises. He had been on hydroxyurea, morphine despite which he has had debilitating sickle pain crises, He also had acute chest syndrome a week prior to stem cell transplant. Allogeneic bone marrow stem cell transplant from haplomatched sibling donor was planned for the patient. Donor blood group was B positive while patient blood group was O positive. The HbS was suppressed to less than 30% by three therapeutic phlebotomy sessions of 200ml and repeated transfusion of leucodepleted packed red cells was given [3].

Testing for viral marker and other biochemical parameters was done to assess the suitability of the donor for bone marrow harvesting. Bone marrow harvesting was done without G-CSF priming as he developed breathlessness and fall in saturation after injection Neukine. 900ml of bone marrow was harvested and procedure well tolerated by the donor. During procedure 2 units of packed red cells were transfused to the donor. Blood bank received harvested product for processing to reduce plasma volume and red blood cells. 37ml product was obtained after processing on Hemonetics MCS$^+$. Product contained 1.1 x 10^6 CD34$^+$cells/kg. Dose was insufficient, so peripheral blood stem cell harvesting was planned from donor after giving inj. Plerixafor. 285ml product was harvested and 6.2 x 10^6 CD34$^+$cells/kg were collected. Donor procedure was uneventful.

Patient developed hemoglobinuria and hypertension reaction after stem cell transfusion, managed conservatively with supportive care. On and off abdominal pain managed conservatively. Engraftment for platelets achieved on day 13 (Platelet count >20000/cu). While ANC >500/mm^3 was attained on day 20. Patient discharged on being hemodynamically stable, afebrile, accepting orally well with Hb 10.5g/dl, TLC-3000, Platelet 152000, N-30%, L-55%. Now patient is coming for regular follow up. His recent HPLC report also showed sickle cell trait and blood group also changed B positive that of donor blood group. Bone marrow transplant is still a hope for patients with severe sickle cell disease although it is not perfect because of associated complications. Haploidentical stem cells transplant have been found to be less successful but in our case haploidendical stem cell transplant was successful.

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