Achievement of cure following allogeneic HSCT with Flu-Bu regimen in a patient with severe mycosis fungoides and Sezary Syndrome

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Experience with allogeneic hematopoietic stem cell transplantation (HSCT) in mycosis fungoides/Sezary syndrome (MF/SS) is limited to a small number of case reports and case series [1,2]. The advantage of allogeneic HSCT has been indicated in progressive disease in the review of CIBMTR study groups [3]. A consensus is still not available about the intensity and the content of the conditioning regimen due to the rarity of the disease and heterogeneous patient groups.

We want to present a refractory MF/SS patient who had catastrophic cutaneous findings and successfully managed with allogeneic HSCT with a sub-ablative conditioning regimen. A 25-year-old female patient was admitted with widespread facial erythematous, squamous papules and large plaques. She was learned to have been diagnosed with MF and treated with total body skin electron beam, interferon and low dose methotrexate. Radiologic examination revealed widespread intra-abdominal, iliac and inguinal lymphadenomegaly with 3 cm of short axis in the largest node. White blood cell count was 8600/ml. Peripheral smear exhibited increased mononuclear cells with cerebriform nuclei. Ratio of CD3+, CD4+, CD7 – Sezary cells was 88% in flow cytometry examination. The patient was evaluated as stage IVA1 (T2N2M0B2) MM/SS according to ISCL/EORTC TNMB staging classification.

Partial remission was obtained following 6 months of photoferesis and Bexarotene treatments in the patient who was refractory to CHOP and GEMOX therapies. Allogeneic HSCT was performed with Bu9.6 Flu150 and rabbit anti-thymocyte immune globulin 30 (rATG-F) conditioning regimen from a male unrelated donor. ABO and Rh type antigen between the patient and the donor was compatible. The number of transplanted CD34-positive cells was 6.3x10⁶/kg. Graft versus host disease prophylaxis consisted of cyclosporin and short-course methotrexate. Neutrophil engraftment occurred on day 12 and platelet engraftment on day 14. Chimerism was determined by PCR-based procedures using STR analysis. Chimerism was found as 90% on post-transplant day 30 and 99% on post-transplant day 100 and 180. Cutaneous lesions disappeared and Sezary cells were not detected in peripheral blood on day 100 after HSCT. The patient was in complete hematologic and radiologic remission. Graft versus host disease did not develop and immune suppressive agents were discontinued at 6th month after HSCT. The patient is still being followed up with complete remission in the third year after transplant. Patient satisfaction could be provided as her excessive cutaneous lesions which had led her to be difficultly recognized completely disappeared.

Conditioning regimen is one of the most important prognostic factors of transplant besides age and performance status of the patient, donor characteristics, disease status at the time of transplant. Whether reduced intensity or myeloablative conditioning suitable for allogeneic HSCT is still controversial [4-8]. It should be considered that non-myeloablative regimens may also be effective in T cell lymphomas due to strong graft versus lymphoma effect. CIBMTR data did not reveal a statistically significant difference between myeloablative and reduced intensity conditioning regimens with regard to overall survival, progression-free survival and mortality in 129 patients although the clinicians carry anxiety for transplant-related mortality with myeloablative regimens [4]. Patient outcomes are encouraging as they indicate that ablative doses could be preferred in proper patients.
We have planned a fludarabine busulphan-based conditioning regimen as infection and *graft versus host disease* risk may increase in patients who have skin involvement and whose skin barrier is impaired. We have used busulfan in sub-ablative dose (9.6 mg/kg) considering disease status and performance status of the patient. We have used high dose rATG-F for benefiting from its anti-tumor effect on T cells and for reducing chronic *graft versus host disease* risk [9]. We consider that allogeneic HSCT with ablative or reduced intensity conditioning which does not contain total body irradiation would be proper for treatment of patients diagnosed with MF/SS. Further studies are required about this issue as limited literature data are available.

**References**


