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Oral Abstracts

OP01

Plasma Cell Leukemia. Experience from tertiary referral centers in Abu Dhabi.

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Introduction:

The diagnostic criteria for PCL is well established and includes plasmacytosis of greater than 20% of total white cell count or an absolute plasma cell count of greater than 2 X 109/L in peripheral blood. It is classified as either primary (pPCL) if there is no antecedent history of Multiple Myeloma (MM) or secondary (sPCL) arising as a leukemic event of MM. Here we report our experience with PCL.

Method:

Eight patients were identified with diagnosis of PCL between January 2008 and March 2016.

Result:

The median age was 53.5 years (range 34-77 years), male to female ratio was 5:3. There were four cases of sPCL while the other four patient's had pPCL. The median time for transformation in sPCL was 199 weeks (range 163 to 244 weeks).

All 4 patients with sPCL had received treatment with novel agents including bortezomib and IMIDS (lenalidomide/thalidomide) as part of their myeloma therapy. At the time of transformation two patients were unfit for any therapy while the other two patients was re-induced with bortezomib, lenalidomide and dexamethasone (RVd) with no response. The median survival after diagnosis of sPCL was 5.4 weeks (range 4-11 weeks).

Patients with pPCL were treated with RVd (n=3) and VMP N=1. One patient also had an autologous stem cell transplant. Patients with primary PCL had a better median survival of 37 weeks (range 14 – 133 weeks) with one patient alive and under therapy 14 weeks after diagnosis.

Conclusion:

Plasma cell leukaemia remains a disease with extreme poor prognosis and an overall mortality of 88 per cent in our experience. The difference in survival between pPCL and sPCL is most likely because the sPCL has a more resistant disease due to previous treatment and clonal evolution of their disease.

OP02

Coexisting Sickle cell disease with hereditary elliptocytosis, a case series and review in the literature

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Background and purpose:

Sickle cell disease (SCD) is a relatively common genetic disorder in Saudi Arabia particularly in eastern province with an estimated prevalence in adult population 17% for trait and 1.2 % for SCD. Hereditary elliptocytosis on the other hand is not infrequently diagnosed in same population with unknown prevalence. We report a case series of SCD with coexisting hereditary elliptocytosis to high light the high prevalence in this population.

Method: A case series of 4 cases with coexistent hereditary elliptocytosis and SCD found during blood film examination in patients visiting secondary care hospital from May 2015 to October 2016. Review to similar cases in the literature was done.

Results: Four cases of SCD found to have coexistent hereditary elliptocytosis. One male 44 years old and three female 49, 36 and 21 years old. With Hb level 9.6, 8.4, 8.7 and 9.7g/dl respectively. Their Hb electrophoresis were (Hb S: 86.3 %, Hb F: 11.2%, Hb A2: 2.5%), (Hb S: 78.4%, HbF: 19.1 %, HbA2: 2.5%), (Hb S: 89.9 %, Hb F: 7.6%, HbA2: 2.5 %) and (Hb S: 81.0%, Hb F: 16.5, %, Hb A2: 2.5 %) in the same order. Iron profile was normal in all cases. Two cases presented to the emergency with crisis, one during routine hematology clinic follow up and the forth with cytopenia secondary to hypersplenisim.

Discussion:

SCD and hereditary elliptocytosis are relatively common disorders in this part of the world. Coexistence of both diseases will not be unexpected in such area, however there is a paucity of similar cases reported in the literature. We expect that coexistent of both disease will eventually affect the course of the disease as both result in shortened RBC survival. Population of eastern province in Saudi Arabia will be the most suitable subject to study the diseases interaction.

OP03

Failure versus Victory: Haploidentical Bone Marrow Transplant In a Developing Country

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Background and Purpose: Many patients and their families with haematological diseases failed to find a fully Human leukocyte antigen (HLA) matched sibling donor for Allogeneic BMT for whom Haploidentical Bone marrow transplant (BMT) is a life saving procedure, in our country haploidentical related donors as allograft source is the only option for these patients. Graft rejection, severe Graft versus host disease (GVHD) and non relapsed mortality are the significant risks associated with haploidentical BMT which are the manifestations of excessive alloreactivity by host and donor T cells

Methods: We have taken up the challenge and pioneered haploidentical related BMT. This program was started from 2004. We performed 21 haploidentical BMT from 2004-2016 at National Institute of Blood Disease & Bone Marrow Transplantation. Diagnoses included leukemia (n=8), thalsasseamia (n=6), Severe combined immunodeficiency disorder (n=3) Fanconi's anemia (n=2), severe aplastic anemia (n=1) and Gauchers Disease (n=1)

Results: Mean age of patients was 13.54years (±13.67) with male predominace. Mean neutrophil engraftment was achieved in 13.41days and platelets were engrafted in 19.08days. Transplant related mortality (TRM) was 47% (N = 10),amongst which leukemic (5/8,62.5%) thalassaemic(1/6,16.6%),SCID(1/4,25%)and 100% in Severe aplastic(1/1) and Fanconi's anemia(2/2,100%) Causes of these deaths were mainly relapse, sepsis, Graft rejection and GVHD. Overall survival rate in our allogeneic patients was 52%.

Conclusion: Haploidentical BMT is the only hope for those patients lacking a full matched donor in hematological malignancies, bone marrow failure and immunodeficiency disorder. Moreover our results are also comparable with global published reports.

OP04

hOCT1 gene polymorphism M420del is associated with decreased response to imatinib in CML patients & its effect is counteracted by M408V polymorphism

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Background and purpose : Human organic cation transporter1 (hOCT1,SLC22A1),an influx transporter,is responsible for the uptake of Imatinib into chronic myelod leukemia (CML)cells. Some patients fail to achieve optimal molecular response to Imatinib, defined as major molecular response (MMR) i.e. BCR-ABL 1 ≤ 0.1% within 12 months of therapy. Variation in clinical response to Imatinib has been observed with two nonsynonymous SNPs in hOCT1 gene ,namely M420del and M408V in some populations.

Methodology: 30 newly diagnosed BCR-ABL positive CML patients in chronic phase,and 30 healthy control subjects, all ethnic Indians ,were recruited in the study. M420del and M408V SNPs were examined by allele specific PCR(AS-PCR) in DNA from PBMCs.After initiation of imatinib therapy ,hematological response was monitored at regular intervals ,and molecular response (BCR-ABL1/ABL1 ratio) assessed after 6 or 12 months .

Results and Discussion :Minor allele frequencies for M420del were 0.18 and 0.1 in CML patients and controls ; for M408V 0.4 and 0.27 respectively, closely paralleling those reported in western population.

No significant association between different genotypes of M420del and M408V was observed with either time to achieve complete hematological response (CHR) (p= 0.341 for both SNPs),or presence of optimal/sub-optimal molecular responses(p=0.125 ,0.629 for M420del and M408V respectively).

To analyze the combined effect of these two SNPs , CML cases were divided into 4 groups.Patients with mutant (homo / heterozygous) M420del and wild type homozygous M408V ,failed to achieve an optimal molecular response to imatinib, unlike those with mutant genotypes (homo / heterozygous) for both SNPs (p=0.02).

Conclusions : Mutant M420del allele may be linked to poor outcome of imatinib treatment in CML,however simultaneous presence of mutant M408V allele appears to circumvent this effect.These SNPs in hOCT1 gene occur at reasonable frequencies in Indian population, to be of clinical interest as predictors of response to imatinib in CML.

OP05

Potential role of vitamin E supplementation in enhancing blood hemoglobin levels in apparently health mildly anemic Pakistani adults

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Background and Purpose

Mild anemia in apparently healthy people is common in the general population in developing countries. It is often left untreated leading to significant morbidity. Previous studies have shown that treatment with vitamin E results in increased colony-forming-units of erythroid progenitors and inhibition of premature erythrocyte lysis. The objective of this study was to determine if vitamin E supplementation could enhance hemoglobin levels in mildly anemic apparently healthy adults in general population of Karachi, Pakistan.

Methodology

In a, placebo-controlled intervention trial, 124 mildly anemic adult males and non-pregnant females (18-45 years), recruited and screened through General Practitioners' clinics and from the personnel of Aga Khan University, Karachi, were randomized into intervention (n= 82) and control (n= 42) groups. Each subject was asked to take one 400 mg vitamin E capsule in intervention group and a placebo capsule in control group daily for three months. Fasting blood samples obtained from each participant at baseline and at end-line were tested for determination of hemoglobin levels and serum concentrations of ferritin, soluble transferrin receptor (sTfR), folate, vitamin B12, erythropoietin, vitamin E, LDL-cholesterol, HDL-cholesterol, glucose and creatinine. The blood/serum levels of these parameters were compared and analyzed statistically using repeated measures ANOVA and multiple linear regression.

Results

There was a significant increase in mean concentrations of both vitamin E and hemoglobin ($P = 0.045$ and $P = 0.049$, respectively) after three months of vitamin E supplementation in intervention group as compared to the control group. The adjusted regression coefficients (ß) and standard error [SE (ß)] of the significant predictors of post-supplemental blood hemoglobin levels were serum concentrations of vitamin E (0.983[0.095]), sTfR (-0.06[0.02]), baseline hemoglobin levels (0.768[0.077]) and gender (-0.656[0.244]).

Conclusion

A positive association between vitamin E supplementation and increased blood hemoglobin levels was found in mildly anemic Pakistani adults.

OP06

Impact of Nutrition on Non-Relapse Mortality and Acute Graft Versus Host Disease during Allogeneic Haematopoietic Cell Transplantation for Haematological Malignancies.

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Background: Allogeneic haematopoietic cell transplantation (HCT) is often associated with poor oral intake. Although it seems obvious that optimal nutrition may improve outcomes, there are no direct data supporting this. It is also unclear whether artificial nutrition support (ANS) should be provided as enteral tube feeding or parenteral nutrition (PN).

Methodology: We analysed day-100 non-relapse mortality (NRM) and incidence and severity of acute graft-versus-host disease (GvHD) according to both route and adequacy of nutritional intake, together with other known prognostic factors. A total of 484 patients who underwent HCT for haematological malignancy at a single institution between 2000 and 2014 were included. ANS was initiated in 228 (47%) patients who met pre-defined feeding criteria.

Results and Discussion: Multivariate analyses showed significant associations of NRM with age > 50 years, previous autograft and positive recipient CMV serology. Additionally, hazard ratios (HR) were significantly increased in the PN and inadequate nutrition groups compared to those with adequate enteral intake - adequate PN: HR 3.2; 95% CI 1.7–6.0, inadequate nutrition: HR 4.4; 95% CI 2.4–8.0; both P<0.001. There were increased incidences of gastrointestinal GvHD of any stage and acute GvHD > grade 1 in patients who received PN (HR 2.0; 95% CI 1.2–3.3; P=0.006, and HR 1.8; 95% CI 1.1–3.0; P=0.018, respectively), but not in those with inadequate nutrition compared to adequate enteral nutrition (EN).

Conclusion: In conclusion the data show that adequate nutrition during allogeneic HCT is associated with improved non-relapsed mortality at 100 days. Adequate EN is associated with significantly better results for this outcome than adequate PN. Furthermore adequate EN, predominantly via oral intake may be associated with lower incidence of overall and gut GvHD when compared to PN, perhaps because of its ability to maintain gut mucosal integrity and support the gastrointestinal tract environment, including gut microflora.