Successful treatment with Extracorporeal Photopheresis of steroid and antibody therapy resistant Cutaneous GvHD following isolated small bowel transplantation in a 5 yr old girl

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Background

- Graft versus host disease (GVHD) following small bowel transplantation (SBT) occurs in about 5-15% % cases [1,2]
- It has a significant impact on transplant related mortality and morbidity particularly in context of a non cancerous setting[3,4]
- In this report we describe the response of steroid and antibody therapy resistant GvHD post SBT to Extracorporeal Photopheresis (ECP)

Clinical report

TRANSPLANT

- A five year old girl underwent ABO matched isolated SBT for microvillus inclusion disease in January 2012.
- GvHD prophylaxis was in form of 6 doses of Basiliximab (20mg) and she was maintained post transplant on Tacrolimus and Prednisolone.

POST TRANSPLANT COURSE

- 8 weeks post SBT : widespread macular rash with hyperpigmented lesions.
- Biopsy : skin Grade III-IV skin GvHD was made. Gut biopsy showed mixed chimerism but no evidence of GvHD.

TREATMENT FOR GvHD

- Methylprednisolone 1mk/kg initially. Lack of response to multiple doses of Infliximab and Basiliximab.
- Mesenchymal stem cells : Total of 4 doses .Following an initial transient response the rash worsened in 2 weeks following the last dose.
- Developed EBV viremia but no evidence of progression to PTLD

EXTRACORPOREAL PHOTOPHERESIS

- Referred at 7 months post SBT.
- On Prednisolone 0.5mg/kg and oral Tacrolimus.
- Cutaneous GvHD affected 70% of body surface area (BSA) with heavily pigmented, lichenoid punctate lesions with intense indurated and exfoliated erythema of palms and soles and severe pruritus. No other organs appeared to be involved.
- Commenced on weekly paired ECP treatment delivered via a femoral catheter with blood priming.
- No alterations to immune suppression were made in the first 6 weeks.
- Week 6 : reduction in erythema to 30 % BSA with particular improvement in the hands and soles. She was commenced on a slow steroid taper at this point and the treatment was given every 10 days.
- Week 12: Prednisolone 0.25 mg/kg with level adjusted Tacrolimus. There is continuing response to ECP with erythema generalised reduction in erythema..
- The pruritus has regressed with no intercurrent infections and stable EBV titres.
- The steroid taper has not been associated with flare up GVHD .

Conclusion

This case report highlights that fact that ECP is a valuable therapeutic intervention in cases of steroid and antibody refractory GvHD

References

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