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Recurrent Post-transplant lymphoproliferative disorder in paediatric heart transplant recipients

Logeswaran T., Behnke-Hall K., Rueblinger L., Bauer J., Thul J., Schranz D.
University hospital giessen paediatric heart center

Introduction: Survival after paediatric heart transplantation (HTX) has improved significantly in the last decades. The long-term outcome is affected by morbidities mostly caused by required immunosuppressive therapy. Post-transplantation lymphoproliferative disorder belongs to the major cause of morbidity. We report about recurrent PTLD in paediatric heart transplant recipients medicated in our institution.

Method: We analyzed the time course of disease, Epstein-Barr Virus (EBV)-status, immunosuppression, involved organs and mortality.

Results: From 1988 till 2010, 156 patients were treated after paediatric heart HTX in our institution. Seventeen patients (11%) suffered from a malignancy. Seven of them (64%) had more than one episode. The group with recurrent PTLD developed the first malignancy on average seven years after HTX (range: 2-13 years). Mean time of recurrence was three years (range: 1-11 years). Most common symptoms were lymph node swelling in six of seven patients (85%). Only two patients had an extra nodal manifestation. Histology of the first malignancy was polymorphic (57%, four of seven) and Hodgkin-like lymphoma in two patients (29%). Second malignancy showed a change in histology: Six of seven patients (85%) had extra nodal manifestations; most common site was the gastrointestinal tract (70%, five of seven). Histology was monomorphic in four of seven cases (57%) and showed Hodgkin-like lymphoma in two cases. Most cases were of B-Cell origin and all patients were Epstein-Barr virus positive. Furthermore, all patients had a CNI-treatment since transplantation and an initial treatment with Azathioprin and were switched to a monotherapy after diagnosis. Treatment of malignancy consisted in CD-20-antibody therapy or chemotherapy (depending on histology). One patient died.

Conclusion: About 11% of paediatric heart-transplant recipients developed PTLD; a high percentage having a relapse. Malignancy was almost always of B-cell lineage and driven by Epstein-Barr virus. Because of high morbidity and mortality, strategies to improve the early diagnosis and the therapy for PTLD are needed.