Intestinal transplantation in children requires close follow-up, including endoscopic monitoring of the transplanted organ via the temporary stoma and/or anus with biopsies taken and reviewed. We present a case of post-transplant lymphoproliferative disorder (PTLD) diagnosed less than 1 month after transplantation. PTLD is a common life-threatening complication after intestinal transplantation, occurring in 13.5% of pediatric cases, and is mostly related to Epstein-Barr virus (EBV) [1, 2].

A 5-year-old boy presented with intestinal failure secondary to microvillus inclusion disease. He received an isolated intestinal allograft combined with a proximal colonic allograft. To monitor for rejection and inflammation, colonoscopy and endoscopic review through the stoma were performed twice a week in the first 2 weeks and once a week after that.

Lesions were detected 26 days after transplantation (Fig. 1a). Microscopy of the transplanted colon and the host colon revealed a polymorphous lymphocytic infiltrate in the lamina propria, non-tumor-forming (Fig. 1b). This PTLD consisted of CD20- and CD79a-positive B cells that harbored EBV-related small RNAs (EBERs) as determined by in situ hybridization (Fig. 1c).

The tacrolimus dose was lowered. However, 6 days later endoscopic review showed that the lesions had grown and were also present in the donor proximal ileum (Fig. 2a). Reduction of immunosuppressive therapy and administration of a monoclonal antibody directed against the B-cell receptor CD20 (rituximab) [3, 4] induced immediate regression of the lymphomas and complete remission of the disorder within 3 months after the first dose.

Six months after transplantation there was an acute episode of therapy-resistant rejection, which needed graft exploration and excision. The patient has been relisted for combined intestinal and liver transplantation. He is awaiting the retransplantation.

plantation at home in a clinically stable condition. This case represents the earliest presentation of intestinal PTLD found during routine endoscopic surveillance.

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References


Bibliography

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