Case Report of Successful Liver Transplantation in a Patient with Acute-on-chronic Liver Failure from Erythropoietic Protoporphyria

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Erythropoietic protoporphyria (EPP) is an inherited disorder of the heme metabolic pathway characterized by accumulation of protoporphyrin in blood, erythrocytes and tissues, and cutaneous manifestations of photosensitivity. Since the major risk in EPP patients is liver disease, regular follow-up for hepatic involvement is essential. At last, hepatic and bone marrow transplantation should be considered as a treatment for most severe cases with hepatic involvement. Herein an EPP patient requiring liver transplantation was described. A 31-year-old male had suffered from photosensitivity since his young age. EPP and concurrent liver cirrhosis were confirmed by skin and liver biopsy 2 years before. Liver cirrhosis progressed leading to acute-on-chronic liver failure with a MELD score of 39. He underwent urgent deceased-donor whole liver transplantation. During laparotomy, the viscera was protected from light source by using a wave length-adjusted filter film since exposure to intense light was known to be a risk factor of bowel perforation. Prolonged continuous renal replacement therapy was necessary to cope with acute renal failure from hepato-renal syndrome. Total porphyrin measurement using blood and urine samples showed negative findings. Immunosuppression was done through combination of cyclosporine, mycophenolate and methylprednisolone. Cyclosporine was a well-known safe immunosuppressive drug for porphyria, which is enlisted as a safe drugs at Cardiff and the Nordic drug database (http://www.drugs-porphyria.org), and other porphyrigenic drugs were avoided. Liver graft function recovered uneventfully, but his hospitalization was prolonged to 3 months due to slow recovery of general condition. The late half of the hospitalization period was spent for rehabilitation treatment enabling standalone walking. This was the first case of EPP in our series of 2,500 adult liver transplants. Bone marrow transplantation will be anticipated if EPP progresses leading to serious clinical manifestations.