

[NM-273] Anesthetic Considerations For Orthotopic Liver Transplantation In A Patient With Multifocal Lymphangioendotheliomatosis With Thrombocytopenia

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Multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT) is a rare complex vascular disorder characterized by diffuse discrete cutaneous and gastrointestinal tract lesions associated with thrombocytopenia [1]. To date, 20 cases of MLT have been reported in the literature. Patients often present with recurrent episodes of life threatening gastrointestinal bleeding in infancy [2]. In addition to the skin and GI tract, multiple organs can be involved in MLT including: lung, muscle, bone, kidney, liver, spleen and CNS [3]. MLT patients may require anesthesia for upper endoscopies for hemostasis management, exploratory laparotomies, MRI sedation, biopsies, and venous access. However, there currently lacks discussion in the literature of anesthetic considerations for patients with MLT. Case Report: This case report discusses a 9 year old 26 kg female with MLT who presented for orthotopic liver transplantation (OLT). The patient had end stage liver disease secondary to MLT involvement of the liver, portal hypertension, esophageal varices, and hepatorenal syndrome. The patient had an uneventful rapid sequence induction. The intraoperative course was complicated by large volume fluid shifts, hemodynamic instability requiring epinephrine and dopamine infusions, and reperfusion was complicated by hyperkalemic arrest requiring chest compressions. The patient was successfully resuscitated and the remainder of the OLT was uneventful. Three weeks later, the patient presented for transjugular liver biopsy for persistent chylous ascites and elevated liver enzymes. Her post-operative course was further complicated by persistent bilateral pleural effusions and a moderate size pericardial effusion without evidence of cardiac tamponade. To avoid positive pressure ventilation, the transjugular liver biopsy was done with supplemental oxygen, intermittent boluses of ketamine and a dexmedetomidine infusion. Anesthetic management of MLT may present major perioperative challenges given the variability of multiorgan involvement. Discussion: Anesthetic considerations of a patient with MLT include the need for perioperative red blood cell, platelet and fresh frozen plasma transfusion. Transfused platelets may have a shortened half life due to platelet trapping in the abnormal dilated vascular channels of the lesions [2]. The fact that OLT has not been previously described in a patient with MLT makes this case unique. The long term prognosis of the patient is unknown and the likelihood of MLT recurrence in the transplanted liver is also uncertain. This case highlights the complexity of MLT. Information sharing among subspecialties is crucial to further our understanding of this rare clinical entity.

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