[NM-192] Controlled drowning in a patient with alveolar proteinosis

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Pulmonary alveolar proteinosis (PAP) is an extremely rare life-threatening condition in which high levels of surfactant prevent normal pulmonary physiology. Although pathogenesis is unclear, it can occur in immunodeficiency. We describe a case of PAP treated with whole lung lavage (WLL) for refractory alveolar proteinosis. Whole lung lavage for PAP removes the lipoproteinaceous material impairing normal oxygenation and ventilation. Unfortunately less invasive treatments have little success.

A 13 year-old 33 kilogram male presented in severe and prolonged respiratory distress. His history is significant for a remote history of heart transplant for myocarditis-induced dilated cardiomyopathy and subsequent lymphoproliferative disease. Lung biopsy revealed the diagnosis of PAP. Despite exhaustive noninvasive therapies, he had worsening secretion burden and hypoxemia, therefore WLL was planned. Our patient’s trachea was measured on chest radiograph and a 26F DLT was planned. Following preoxygenation and induction saturations immediately declined to the low 70s but were able to be recovered to baseline after intubation and correct positioning of the DLT. Tube position was verified by fiberoptic scope, auscultation, fluoroscopy and leak test prior to and following every third cycle of 400 ml lavage. The leak test consisted of applying positive pressure to the tracheal side while bronchial side was placed underwater to evaluate for bubbles. The left lung was lavaged with a total of 9 liters of normal saline, which showed a dramatic improvement in the clarity of liquid retrieved from the lung over time. The DLT was then exchanged for a single lumen ETT and the patient was taken to the ICU for recovery. Right lung lavage was performed by the same technique but was delayed a week due to the patient developing pneumonia. The subsequent right lung lavage was not tolerated as well due to desaturations occurring during each lavage but promptly returning to baseline when two lung ventilation was resumed. However 12 cycles were completed again with dramatic clearing of the retrieved liquid and the patient was again returned to the ICU with a single lumen ETT for recovery.

Lung isolation is absolutely indicated and the most critical part of the anesthetic technique. This can be very complicated in pediatric practice the younger the patient is. Several very creative solutions have been published. Due to the uncertainty of how patients will tolerate WLL we had ECMO on standby and this should always be considered in such cases. Arterial cannulation is essential for blood gas determination during the procedure. Consideration should also be given that alveolar ventilation is severely compromised and volatile anesthetic uptake may not be reliable during lung lavage. We recommend Bispectral index monitoring and intravenous anesthesia as an important part of the anesthetic technique.

References:

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