Case Report: Extremely Rapid Deterioration in a Child with Plastic Bronchitis: What’s Changed in the Last Century?
Elena H. Cho, MD and Michael J. Mulick, DO
Department of Anesthesiology and Critical Care Medicine, Children’s Hospital Los Angeles

Abstract:
Plastic bronchitis has been described in literature for over 100 years with an incidence reported to be somewhere between 4 to 14% with very high mortality. The authors describe a case of plastic bronchitis in a fontan patient with an insidious presentation and an extremely rapid deterioration. Effective therapy requires early diagnosis and a multidisciplinary protocol that emphasizes early bronchoscopic extraction. Other methods such as lymphatic embolization and high frequency jet ventilation should be discussed early and implemented as soon as possible. Adequate parental preparation is imperative.

Case:
An 8 year old male with a history of Fontan palliation for single ventricle physiology presented to the emergency room with respiratory distress. He had a mildly productive cough and was admitted with a presumed diagnosis of pneumonia. His condition worsened over several days and was taken to the operating room for a bronchoscopy and removal of thick gelatinous material that was confirmed by pathology to be plastic bronchitis. Over the next 2 days he required intubation, vasopressors, and nebulized tissue plasminogen activator. The patient expired before subsequent bronchoscopy and any other therapy could be attempted. The time from diagnosis to expiration was less than 60 hours.

Discussion:
Plastic bronchitis has been described in medical literature for over 100 years and etiologies are usually of an inflammatory type such as asthma, pulmonary infections, and cystic fibrosis. It is also seen in children following congenital cardiac surgery. It is rare and life-threatening, and characterized by large, pale, and rubbery bronchial casts that develop in the tracheobronchial tree which causes complete airway obstruction, multiple end organ failure, and death.

Endobronchial lymphatic leakage has been suspected to play a role in cast formation and some institutions have described successful management with lymphatic imaging and embolization of peribronchial lymphatic channels. High pulmonary venous pressures have also been implicated in causing an abnormal response of respiratory epithelium resulting in excess mucous production. Aerosolized tissue plasminogen activator and the use of high frequency jet ventilation have been described as effective modalities for destroying casts.

Plastic bronchitis remains highly lethal as it is difficult to diagnose early as symptoms mimic other respiratory ailments, requiring practitioners to have a high index of suspicion in children with history of congenital cardiac surgery. Even after diagnosis, it remains extremely difficult to manage, and until recently the only effective modality has been bronchoscopic extraction. The options for treatment diminish once patients deteriorate as profound hemodynamic instability can occur rapidly. An emergent bronchoscopy and or high frequency jet ventilation and lymphatic embolization should be considered and implemented early. This case highlights the need for multidisciplinary discussions, parental preparation, protocol implementation, and early intervention to improve mortality in these patients.

References:
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