Plastic bronchitis is an unusual condition, in which extensive endobronchial casts arborize throughout the lung to cause coughing, wheezing, expectoration of mucoid plugs or casts, hemoptysis, respiratory distress or even death. It differs from ordinary mucus plugging due to its cohesiveness and notoriously difficult removal via bronchoscopy. It can occur in patients with asthma, bronchopulmonary aspergillosis, with cystic fibrosis and, less commonly, after surgery for congenital heart disease (1-3). Specific to post-operative patients, this process is a complication primarily seen with the Fontan procedure, which is performed for children with congenital single ventricle heart disease in order to divert systemic venous return into the pulmonary circulation.

CASE PRESENTATION

A six-year-old male with a history of double-inlet left ventricle, great vessel transposition and an interrupted aortic arch who previously underwent a Norwood procedure, arch reconstruction and bidirectional Glenn in infancy presented for a fenestrated Fontan procedure. He was admitted to the pediatric cardiac intensive care unit (PCICU) post-operatively. His course was complicated by increased work of breathing on hospital day 16, and chest radiograph revealed complete right lung atelectasis and pleural effusion. Endotracheal intubation became necessary for persistent hypoxia. Bedside bronchoscopy demonstrated tenacious plugging.

Further respiratory decompensation required emergent intervention by Pediatric Otolaryngology in the operating room. The endotracheal tube was removed, and direct laryngoscopy revealed normal anatomy to the level of the subglottis. Advancement of a ventilating bronchoscope to the carina revealed a patent left mainstem bronchus and a completely obstructed right mainstem bronchus (Figure 1). The grasping optical forceps were passed multiple times to remove large soft casts until the endobronchial tree was cleared (Figure 2). His oxygen saturations increasingly improved throughout the case, and right-sided chest rise was eventually observed. He was reintubated and returned to the PCICU for ongoing cardiopulmonary management. He was treated with dornase alph and alteplase nebulizations, though he required an additional bronchoscopic intervention for cast removal during his hospitalization.

DISCUSSION

Galen (131-200 AD) first hypothesized that bronchial casts were expectorated pulmonary blood vessels. Morgagni thought these structures consisted of inspissated mucus. At the turn of the 20th century, plastic bronchitis was thought to be caused by asthma or infection due to tuberculosis or even adherent pericarditis (1, 2). In 1997, Seear et al categorized bronchial casts based on histology, and this classification system has persisted through the literature (3). The inflammatory type I casts contain fibrin with a dense eosinophilic infiltrate, are associated with bronchopulmonary disorders and are more amenable to medical management. The acellular type II casts consist predominantly of mucin, are associated with post-operative congenital heart disease and are much more likely to require repeated urgent bronchoscopy.

There are several theories on how the casts develop in post-operative congenital heart disease patients (2).

REFERENCES