LETTER TO EDITOR (VIEWERS CHOICE)

PLASTIC BRONCHITIS: BRONCHIAL CASTS MIMICKING FOREIGN BODY ASPIRATION

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A 3-year old girl presented with history of coughing up a piece of "chicken meat". The patient's sister also reported that the patient had coughed up "noodles" the night before. The child was slightly cyanotic during the coughing episode that brought up the material, but was not in distress. This patient was born with a hypoplastic left heart which required multiple operations beginning at 2 weeks of life. She had a Sano-Norwood procedure followed by a Bidirectional Glenn procedure at 5 months of age. The patient had an episode of chylothorax at 12 months of age, which was thought to be a complication of her cardiac surgeries and elevated intrathoracic pressures. Since that time, the child had been on a low-fat diet. Subsequently, the patient underwent an extrinsic non-fenestrated completion Fontan procedure 4 months prior to her presentation. Despite having palliative cardiac operations, the patient was doing well at home without baseline cyanosis. However, she was admitted to our hospital 1 month prior for a presumed pneumonia which was treated with antibiotics. The patient had a baseline oxygen saturation of 92% on room air following treatment and upon discharge. At the time of evaluation, the patient was not cyanotic on room air, had an oxygen saturation in the low 90%'s, dropping to the 80%'s while she coughed intermittently. She was speaking and playing normally. Her head and neck examination was unremarkable, but she did have diminished breath sounds bilaterally, worse on the right side. Her chest x-ray showed mild pulmonary edema without evidence of hyperinflation on decubitus films. Nor was there evidence of a radio-opaque foreign body. A rigid bronchoscopy revealed a large mucus cast obstructing the bilateral main stem bronchi at the level of the carina. As we extracted the mass, there appeared to be arborization of the mass down the secondary and tertiary bronchi. (Figure 1) Pathologic examination revealed an amorphous, acellular mucus cast composed primarily of mucin with a few scattered foci of inflammatory cells. There was no bacterial infection. The patient was transferred to the intensive care unit (ICU), intubated and required a second removal of the plastic casts 4 days later due to rapid re-accumulation. She was successfully extubated afterwards and thus far has not had a recurrence of plastic bronchitis.

Plastic bronchitis is a rare condition of bronchial obstruction with a mucus cast that can be life threatening. (1) It is described in adults and children, although the incidence in adults is exceedingly rare. The clinical presentation varies, with the most commonly described history including respiratory distress while coughing up foreign appearing material; specifically "chicken meat" or "noodles." (2) Seear et al (3) described a classification of these mucus casts as Type I (cellular) or Type II (acellular). These casts are different from ordinary mucus casts because of their consistency and cohesive nature that results in the casts taking the shape of the bronchi in which they are formed. Type I casts occur in the setting of pulmonary

Figure 1: Arborized mass

inflammatory disease such as pneumonia, asthma, bronchitis, allergic bronchopulmonary aspergillosis, cystic fibrosis, and acute chest syndrome in patients with sickle cell disease. Histologically, Type I casts show primarily fibrin with little mucin and have an eosinophilic infiltrate. Type II casts occur in patients after surgery for congenital cardiac defects, especially those with a Fontan physiology. This is believed to arise because of high central venous, poor ventricular function, and bronchial hypersecretion. Some have also suggested that the pathophysiology is linked to the development of lymphatic abnormalities and endobronchial chyle leakage as a result of surgical trauma, adhesions or elevated central venous pressures. Type II casts are acellular and composed mainly of mucin with little or no inflammatory infiltrate.

Our patient has a Fontan circuit and her pathology was characterized as a Type II plastic cast. Most cases of Type II casts occur between 1 to 3 years after a completion Fontan procedure. Our patient's preceding pneumonia likely accelerated the cast process, causing this occurrence just 4 months after her operation. In addition, her previous history of chylothorax signals an alteration of her lymphatic circulation that may have ominously hastened her development of plastic bronchitis.

Because of the acute and vague presentation, mortality from plastic bronchitis is reported to be as high as 29%. (1) Most fatalities reported are due to asphyxiation caused by airway obstruction. Flexible bronchoscopy has been used for diagnosis, but for effective clearance of the casts, the otolaryngologist must often perform a rigid bronchoscopy. (4) Use of optical forceps and flexible suction can be very helpful in these cases. The casts are extremely pliable and difficult to extract with forceps alone. However, irrigation with normal saline causes the casts to swell and thus become easier to grasp and remove. The long term prognosis for this disease is poor. After acute extraction of the casts via rigid bronchoscopy, the

casts tend to re-occur unless the underlying problem is resolved. (4) In the case of inflammatory casts, long term antibiotic therapy and corticosteroids (inhaled and systemic) have been tried. Macrolide antibiotics, which are believed to have anti-inflammatory and mucoregulatory properties have also been used. (2,5)

Lytic therapies include chest physiotherapy, mucolytic inhalation with potassium iodide (SSKI) or acetylcysteine. (2) Recombinant human DNase which hydrolyses extracellular DNA has been reported to be effective in assisting extraction of plastic casts. (6) Finally, casts that have large fibrin content have been treated with fibrinolytics such as heparin, urokinase, and tissue plasminogen activator. (7)

For Type II casts, therapy is aimed at improving cardiac function and decreasing venous pressure and intrathoracic lymphatic pressure. Heparinization and adherence to a low-fat diet are frequently employed. Fenestration of the Fontan baffle may also decrease pulmonary arterial pressure. Finally, thoracic duct ligation has been used to reduce lymphatic pressure and prevent chylothorax. (8)

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