Persistent Plastic Bronchitis in a Child after Cardiac Surgery

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Plastic bronchitis is a rare clinical entity that occurs as a complication of asthma, allergy, respiratory tract infections, cystic fibrosis, sickle cell disease, pericarditis and congenital heart defects [1,2]. Most cases related to cardiac defects are reported to have occurred after corrective surgery, mainly the Fontan procedure [1–3]. The Fontan procedure creates a direct connection between the systemic venous circulation, most frequently the superior and inferior vena cavas, and the pulmonary artery. Plastic bronchitis presents with severe respiratory distress and is usually diagnosed when white casts are expectorated or detected by bronchoscopy [1]. We describe a patient who developed fatal plastic bronchitis after undergoing the Fontan procedure for repair of tricuspid atresia.

Patient Description

A 6 year old girl was born with a cyanotic heart defect that was neither diagnosed nor treated in her country of birth. Until the age of 3 years she suffered from chronic hypoxia, which was the probable cause of her mild psychomotor retardation. At age 3, immediately after she immigrated to Israel, she was diagnosed as suffering from tricuspid atresia with a single ventricle. During the next 3 years she underwent a series of operations: a right modified Blalock-Taussig procedure, placement of an aortopulmonary shunt, placement of a bidirectional Glenn shunt, and finally, the Fontan procedure. After her last surgery she developed pneumonia in the right lung, atelectasis of the left lung, and chylothorax, all of which resolved. She also sustained permanent paralysis of her right diaphragm. Echocardiography revealed normal left ventricle function, mild mitral regurgitation, and good flow through the Fontan conduit. She was discharged home in good condition and treated with diuretics and coumadin.

Two months after being discharged and 3 months after undergoing the Fontan procedure, she arrived at the pediatric emergency room of our hospital with severe dyspnea and cyanosis. According to her parents' report she developed a cough 2 weeks previously. She was afebrile, her respiratory rate was 60 per minute and blood pressure 120/80. Oxygen saturation on room air was 50%, and 88–90% with an oxygen mask. On auscultation, breath sounds were reduced over the right lung field. No clinical signs of heart failure, significant abdominal findings or clubbing were noted. A complete blood count revealed a white blood cell count of 15,600 with 89% neutrophils, and hemoglobin and platelets within the normal range. There was no infiltrate, atelectasis or hyperinflation on the chest X-ray, but the right diaphragm was elevated.

She was hospitalized in the pediatric intensive care unit and initially treated with oxygen, chest physiotherapy, bronchodilators and intravenous antibiotics. The next day her condition improved significantly and she was transferred to the pediatric ward. Four days later her respiratory state worsened and she was returned to the pediatric intensive care unit. Chest X-ray showed partial atelectasis of the right lung. Emergency flexible bronchoscopy demonstrated a cast blocking the entrance to the right main bronchus, which was immedi-

ately removed by rigid bronchoscopy. The cast material was solid mucous in the form of a mold of the bronchial tree [Figure]. Her respiratory status later improved considerably. Cultures taken from the cast were negative. Pathologic examination of the cast revealed a protein exudate mixed with few neutrophils and lymphoid cells. During the same hospital stay there were repeated episodes of respiratory deterioration, each requiring bronchoscopic removal of similar casts from the right bronchial tree. Therapy with inhalation of acetylcysteine and urokinase, as well as with systemic steroids and azithromycin was unsuccessful in preventing further cast formation.

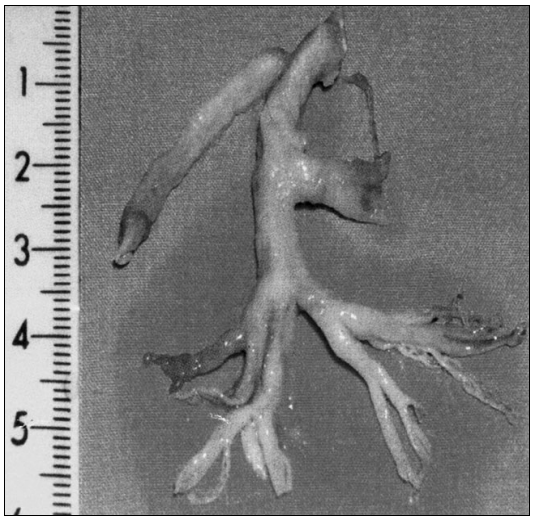
In an effort to improve the effectiveness of her cough and her ability to clear cast material from her right lung, plication of the right diaphragm was performed. This too failed to improve her course. Cardiac catheterization revealed slightly reduced cardiac output and normal pressures in the Fontan circulation.

In the ensuing months she was hospitalized every 2–3 weeks for similar episodes of respiratory distress, all treated by bronchoscopic removal of newly formed casts. Treatment with tissue plasminogen activator inhalation during one hospitalization showed no clear benefit. During her final hospital stay, and 7 months after the Fontan procedure, she developed severe respiratory failure with a room air oxygen saturation of 80%. Rigid bronchoscopy was performed, during which she suffered a cardiopulmonary arrest. Despite all resuscitative efforts, she died a few hours later.

Comment

To the best of our knowledge the incidence of plastic bronchitis is not mentioned in the literature, though it is considered a rare entity with only approximately 50 cases published to date [1–5]. We report here a 6 year old girl born with a congenital cyanotic heart defect, who developed plastic bronchitis 3 months after a Fontan procedure and died 4 months later. Despite treatment with various medications and multiple rigid bronchoscopies for removal of newly formed casts, she succumbed to her disease. We speculate that the large cast caused her severe hypoxia prior to the last bronchoscopy and initiated the rapid deterioration that worsened during the procedure and caused the child's death.

Seear et al. [2] in 1997 reported nine cases of plastic bronchitis and suggested a classification of two types according to the cast composition. Type I refers to inflammatory casts that contain mainly fibrin with a rich inflammatory cell infiltrate; these casts appear in inflammatory lung diseases. Classified as type II are acellular casts composed primarily of mucin, with few inflammatory cells; this type occurs in congenital heart defects after surgical repair. In pulmonary diseases, type I casts are formed by the affected bronchial airways. The mechanism accounting for the overproduction of mucin by the respiratory epithelium in type II cannot be directly explained by the pathophysiologic changes that occur before or after surgical correction of congenital heart defects. Seear and colleagues [2] suggested that the underlying cause is pulmonary venous hypertension; however, Languepin et al. [3] proposed lymphatic leakage in the chest as the cause of cast formation.



Bronchial cast extracted from the patient.

The prognosis is largely determined by the etiology. Brogan and associates [1] found that patients with a congenital cardiac defect had a mortality rate of 29% and a life-threatening events rate of 41%. No deaths or life-threatening events occurred among patients whose underlying disease was asthma or allergy [1]. Languepin et al. [3] described 14 patients who developed plastic bronchitis secondary to congenital heart defects, of whom 43% died [3]. Seear's group [2] claimed that if correction of cardiac dysfunction is impossible, then the prognosis is poor [2].

Treatment of type I disease includes empiric administration of systemic or inhaled steroids, antibiotics, bronchodilators and mucolytics. For type II disease the medical treatment is more problematic. There are a few sporadic reports in the literature of successful treatment of type II plastic bronchitis using a variety of treatment modalities. Inhalation of urokinase or tissue plasminogen activator, and intratracheal instillation of rhDNAse led to improvement in some patients, though their effect on mucin is unclear [1]. Both oral azithromycin and subcutaneous heparin when administered long term proved effective in individual cases [4,5]. In our patient, all medications failed to prevent recurrence of casts.

The purpose of drug therapy is to prevent reproduction of casts. However, once formed, the casts must be removed with a rigid bronchoscope as their stiffness precludes use of a flexible one. Our patient underwent immediate rigid bronchoscopy for each respiratory exacerbation. In most instances, a single solid mucous cast in the form of a mold of the bronchial tree was removed either intact or in smaller pieces. In light of the high rate of treatment failure, for severe cases that develop in children after repair of congenital cardiac defects, cardiopulmonary transplant should be considered in the early stages.

In conclusion, we describe a young girl who developed plastic bronchitis after the Fontan procedure, and despite aggressive management she succumbed from this disease. Unfortunately, the optimal treatment for the prevention of bronchial acellular cast formation has yet to be found. It is important to recognize this potentially fatal complication of cardiac surgery for congenital defects. The pathophysiologic mechanism of this process requires further investigation so that a better prognosis may be achieved.

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