Case Report

A near lethal case of plastic bronchitis following truncal valve replacement

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Abstract. Plastic bronchitis is a condition in which large, rubber-like, bronchial casts develop in the tracheobronchial tree causing airway obstruction. It is an unusual disorder that occurs in various disease states. Most case reports of plastic bronchitis associated with congenital heart disease are in patients palliated with Fontan physiology. We describe a 13-year-old girl with DiGeorge syndrome and truncus arteriosus who underwent an uneventful truncal valve replacement. The child developed plastic bronchitis on postoperative day eight requiring extra-corporeal membrane oxygenation support and daily bronchoscopy to remove bronchial casts. Our patient did not have elevated systemic venous pressures or Fontan physiology. We speculate the etiology of plastic bronchitis may have been surgical trauma to the lymphatic channels surrounding the bronchi and disruption of pleural adhesions. A comprehensive evaluation revealed no specific cause and the diagnosis of idiopathic plastic bronchitis was made.

Keywords: Plastic bronchitis, bronchial casts, congenital heart disease, truncus arteriosus, truncal valve replacement

1. Introduction

Plastic bronchitis is a condition in which large, rubber-like, bronchial casts form in the tracheobronchial tree and cause airway obstruction. The mechanism of plastic bronchitis is unknown and the diagnosis is typically made with the expectoration of casts. Expectoration can be spontaneous but more commonly requires bronchoscopy for diagnosis and removal. Death due to airway obstruction from a cast is not uncommon [1]. The treatment of plastic bronchitis consists of supportive care, therapeutic bronchoscopy, and treatment of underlying disease states [2]. Numerous systemic illnesses are associated with

plastic bronchitis including bronchopulmonary diseases associated with mucus hypersecretion, bronchopulmonary bacterial infections, congenital and acquired cardiac diseases or pulmonary lymphatic abnormalities [3]. Most case reports of plastic bronchitis associated with congenital heart disease are described in patients palliated with Fontan physiology.

In this article we describe a girl with DiGeorge syndrome and truncus arteriosus who underwent an uneventful truncal valve replacement and developed plastic bronchitis on postoperative day eight requiring extra-corporeal membrane oxygenation support, aggressive medical management and daily bronchoscopy.

2. Case report

Our patient is a 13-year-old female with DiGeorge syndrome and truncus arteriosus diagnosed at birth.

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A complete repair of her truncus arteriosus was performed at day of life seven and included patch closure of a ventricular septal defect, separation of the pulmonary arteries from the arterial trunk and establishment of right ventricle to pulmonary artery continuity with a valved pulmonary homograft. Over the years, she has required several additional open cardiac operations for conduit changes and two truncal valve repairs for truncal valve insufficiency. She had tolerated her previous operations relatively well and presented again with a mixed truncal valve stenosis and severe insufficiency. She was asymptomatic from a cardiac standpoint and had no significant limitations in her activity. She had no delays in her growth or cognitive development. Her past medical history is notable for respiratory syncytial virus and pneumonia in infancy requiring hospitalization. Following a conduit change as a young child, she developed subglottic stenosis and had recurrent episodes of stridor treated with racemic epinephrine. She also had a subglottic cyst, which was removed. The patient had recurrent otitis media, ruptured tympanic membranes, as well as significant sinusitis requiring multiple sinus surgeries. A cystic fibrosis workup demonstrated the patient and her mother were carriers.

Preoperative transthoracic echocardiography demonstrated severe truncal valve insufficiency and stenosis with a peak gradient of 134 mmHg and a mean gradient of 86 mmHg. Her systemic left ventricle was dilated and hypertrophied, though maintained a normal shortening fraction. Her right ventricle was of normal size and function. The echocardiographic data was diagnostic and a pre-operative cardiac catheterization was not indicated. Her preoperative chest x-ray and laboratory values were all within normal limits. She was admitted to the hospital and underwent an elective truncal valve replacement using cardiopulmonary bypass and cardioplegic arrest. The operation was uneventful other than the redo nature of her heart condition and extensive pleural adhesions throughout the mediastinum and both pleural spaces. A 23 mm mechanical valve was placed in the truncal position. The previously placed right ventricle to pulmonary artery conduit showed no signs of failure, and was left in situ. The patient was weaned from cardiopulmonary bypass requiring no inotropic or vasoactive support and required no transfusions of packed red blood cells or clotting factors.

The patient was extubated on postoperative day one, was ambulating and receiving standard pulmonary toilet to include mobilization, chest physiotherapy and incentive spirometry. Daily upright chest x-rays continued to demonstrate right, middle and lower lobe atelectasis

despite escalation of therapy. She remained on 2-4 L of oxygen via nasal cannulae and maintained oxygen saturations in the nineties. The patient remained stable from a respiratory standpoint other than a nonproductive cough. Per protocol anticoagulation was started with warfarin for her mechanical valve with a goal international normalized ratio of 2-2.5. On postoperative day eight the patient was ambulating comfortably, tolerating a full diet and remained afebrile although persisted with significant right, middle and lower lobe atelectasis on chest X-ray. A flexible bronchoscopic examination was performed demonstrating a large cast obstructing the right mainstem bronchus. The cast was extensive, extending from the carina to the bronchus intermedius. The cast was mucoid in appearance and could not be dislodged by suctioning. The diagnosis of plastic bronchitis was entertained at this time. Flexible bronchoscopy was aborted with plans for rigid bronchoscopy.

The patient remained stable on nasal cannulae with saturations in the 90's. The following morning, the patient was brought to the operating room for rigid bronchoscopy under general anesthesia. Again, the large obstructing mass was noted in the right mainstem bronchus. During the process of removing the mass, a section fractured and became lodged in the left mainstem bronchus. Profound bradycardia and hemodynamic collapse soon followed requiring cardiopulmonary resuscitation. The child was profoundly cyanotic, unable to ventilate with immeasurable oxygen saturations. Emergent, veno-arterial extracorporeal mechanical circulation was rapidly initiated through the right neck approach cannulating the carotid artery and internal jugular vein. Full flow and support were quickly obtained allowing safe reentry into the airway with rigid bronchoscopy to evacuate the bronchial cast (Fig. 1).

The child returned to the intensive care unit on veno-arterial extra-corporeal membrane oxygenation (ECMO) support and correction of a significant metabolic and respiratory acidosis over the next few hours was achieved. Coumadin was discontinued and systemic heparin was initiated and adjusted to standard ECMO protocols. A transthoracic echocardiogram demonstrated preserved, biventricular function, a well seated truncal valve and good decompression of the right ventricle. While on ECMO her airway management consisted of ventilatory rest settings of spontaneous intermittent mechanical ventilation (fraction of inspired oxygen ratio 40%, positive end-expiratory pressure of 10 cm H₂O, tidal volume of 6 cc/kg, rate of 10 bpm), pulmonary toilet, high dose inhaled steroids, nebulized acetylcysteine and nebulized dornase alfa in addition to daily

Fig. 1. Bronchial cast from right mainstem bronchus removed via rigid bronchoscopy.

bronchoscopic cast removal. Aerosolized tissue plasminogen activator (t-PA), while described in the literature [2], was not a safe option for our patient secondary to recent complex, reoperative, open-heart surgery. With the support of ECMO, the child underwent daily bronchoscopic examinations demonstrating large, densely adherent, bloody clots extending into both mainstem bronchi and down into the secondary bronchi. Despite ongoing anticoagulation, these clots became less dense and more manageable with each examination over the next five days. On postoperative day fourteen, bronchoscopy demonstrated thin, clear secretions with no casts or clots. The patient was weaned from ECMO and supported with pressure regulated mechanical ventilation. She was extubated the following morning and treated with aggressive pulmonary toilet. Her warfarin was reinstituted per protocol and she was discharged home on postoperative day 25 with a clear chest x-ray, on room air. Aggressive pulmonary toilet and high dose inhaled steroids were continued post-discharge. At a two month follow up, the patient remained asymptomatic without expectoration of plugs or casts and had a clear chest x-ray.

3. Discussion

Plastic bronchitis is very rare and characterized by the expectoration of bronchial casts. These casts may be associated with bronchopulmonary diseases of mucus hypersecretion, bronchopulmonary bacterial infections, congenital and acquired cardiac diseases or pulmonary lymphatic abnormalities [3]. Plastic bronchitis occurs in a wide variety of noncardiac conditions, including asthma [4–7], pneumonia [8], lymphangiomatosis [9–11], allergic

bronchopulmonary aspergillosis [12], cystic fibrosis [13], bronchiectasis [7,14], acute chest syndrome associated with sickle cell disease [15], smoke inhalation [16] and idiopathic states [4,5,8,11,17,18]. The casts are of variable sizes and take the shape of the bronchi in which they formed. Seear et al. [4] proposed that bronchial casts might be divided into two distinct clinicopathologic groups. Type I, inflammatory casts, are composed of fibrin with little mucin and have an eosinophilic infiltrate. Inflammatory casts are typically seen in patients who develop plastic bronchitis with underlying bronchial disease such as asthma and cystic fibrosis. Type II, acellular casts, are composed mainly of mucin with little fibrin and no inflammatory infiltrate. Patients with palliated congenital heart disease and plastic bronchitis typically present with type II inflammatory casts. Our patient's casts had histologic features of dense mucin deposition with minimal cellularity suggestive of an acellular process.

Pediatric patients with plastic bronchitis associated with congenital heart disease have been described most frequently in palliated, single-ventricle, Fontan physiology [2–4,7,19–24] (Table 1). Zaccagni et al. [23] reported a case of plastic bronchitis presenting as late as nine years after a patient's Fontan operation. Similarly, a patient five years out from a lateral tunnel Fontan presented with plastic bronchitis and required surgical fenestration of the Fontan and drainage into the left atrium [24]. Other pediatric heart conditions associated with plastic bronchitis include tetralogy of Fallot [3,25], anomalous pulmonary venous return [3] and constrictive pericarditis [26] (Table 1).

The pathophysiology in the Fontan subgroup involves the development of endobronchial lymphatic congestion [3,19]. Pulmonary lymphatic abnormalities may be

Table 1
Summary of patients with plastic bronchitis associated with congenital heart disease

References	Diagnosis	Physiology
3, 4, 7, 20, 22	Tricuspid atresia (n = 5)	Fontan
4	Complex cyanotic CHD (n = 4)	Fontan, BDG, BTS(2)
21	DORV, PA	Fontan
19	PA, IVS, D-TGA, MA, Hypoplastic LV	Fontan
2	Double inlet left ventricle, L-TGA	Fontan
23	HLHS	Fontan
24	Unbalanced AV canal	Fontan
3	ASD, PAPVR	Two ventricle
26	Mitral regurgitation, constrictive pericarditis	Two ventricle
3, 25	Tetralogy of fallot (n = 2)	Two ventricle

ASD = Atrial septal defect; AV = Atrioventricular; BDG = Bidirectional Glenn; BTS = Blalock Taussig shunt; CHD = Congenital heart disease; DORV = Double outlet right ventricle; D-TGA = D-transposition of the great arteries; HLHS = Hypoplastic left heart syndrome; IVS = Intact ventricular septum; LV = Left ventricle; L-TGA = L-transposition of the great arteries; MA = Mitral atresia; PA = Pulmonary atresia; PAPVR = Partial anomalous pulmonary venous return; PV = Pulmonary valve.

congenital or acquired. They may be acquired secondary to elevated central venous pressure commonly found in Fontan physiology or even direct injury to the lymphatic system from intraoperative trauma during the dissection of adhesions [3]. Our patient did not have elevated systemic venous pressures or Fontan physiology. We can only speculate the etiology of plastic bronchitis in this case may have been surgical trauma to the lymphatic channels surrounding the bronchi and disruption of pleural adhesions. A comprehensive evaluation demonstrated no definitive cause and the diagnosis of idiopathic plastic bronchitis was made.

A high index of suspicion is necessary to make the diagnosis of plastic bronchitis if a cast has not yet been demonstrated. In patients who are at risk and develop unexplained wheezing and desaturation, high-resolution computed tomography may be useful as a noninvasive study, although we maintain a low threshold for diagnostic bronchoscopy. Supportive care, therapeutic bronchoscopy and treatment of underlying disease states are the basis for treatment of plastic bronchitis [4,5,8]. Plastic bronchitis is a rare disorder associated with variable disease states, therefore, specific therapeutic options are based primarily on anecdotal experience. Traditional treatments include nebulized acetylcysteine, aerosolized urokinase and tissue plasminogen [2]. Nebulized acetylcysteine, [4,8,18] breaks disulfide bonds in mucoproteins, decreasing the viscosity. A case report of aerosolized urokinase in a five year old patient with Fontan physiology and life-threatening plastic bronchitis has been described [20]. Clinical improvement was noted following the initiation of urokinase therapy in this patient. These authors also

demonstrated that bronchial casts removed from their patient were unchanged with in vitro saline incubation but became soft and friable when incubated with urokinase and dissolved completely when incubated in a t-PA solution. t-PA causes fibrin-enhanced conversion of plasminogen to plasmin, which initiates local fibrinolysis. With close proximity of complex, reoperative, open-heart surgery in our patient, lytic therapy was not ideal. Given the mechanism of these agents, we speculate that patients with inflammatory casts with high fibrin concentration may benefit more from nebulized t-PA than those with acellular casts.

Gossett et al. [27] provided a multicenter study in patients with single ventricle physiology and plastic bronchitis who underwent an orthotopic cardiac transplantation. In patients who survived past 30 days there was complete cessation of cast formation. This study did not investigate if alternative therapies were first applied and therefore was unable to compare current therapies to transplantation as a superior treatment. Thoracic duct ligation has also been described as a surgical option in some cases of plastic bronchitis [28,29].

In summary, plastic bronchitis is an uncommon and under diagnosed complication of congenital cardiac surgery, characterized by recurrent expectoration of large, branching bronchial casts. Our patient emphasizes that plastic bronchitis can present as an acute life-threatening emergency if mechanical obstruction of major airways occurs. The treatment of plastic bronchitis includes acute therapy to aid the removal and expectoration of casts, and specific short- or long-term treatments to address the underlying hypersecretory process. Our patient responded well to nebulized acetylcysteine and dornase

alfa, as well as daily bronchoscopy while on supportive ECMO. While this strategy is not always necessary, it remains a viable option in the severely decompensated patient with life threatening plastic bronchitis.

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