

A Case of Plastic Bronchitis Presenting 9 Years After Fontan

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Abstract A 15-year-old boy with hypoplastic left heart syndrome experienced plastic bronchitis 9 years after completion of a nonfenestrated lateral tunnel Fontan. Despite cardiac catheterization with coil embolization of collateral vessels and initiation of a pulmonary toilet regimen, including aerosolized tissue plasminogen activator, he continued to expectorate large acellular-mucinous casts. Finally, after optimization of cardiac function with the addition of carvedilol, the expectorated casts decreased in number. This report reviews pathophysiology of plastic bronchitis cast formation and therapy in the context of this late presentation after Fontan.

Keywords Cast bronchitis · Fontan physiology · Plastic bronchitis

Plastic bronchitis, or cast bronchitis, is a rare disease characterized by the formation of large gelatinous or rigid endobronchial airway casts [2, 5]. Patients either expectorate casts spontaneously, coughing up large impressions of their tracheobronchial tree, or may require bronchoscopy for removal of casts to avert impending respiratory arrest due to airway obstruction.

Plastic bronchitis has been associated with a variety of diseases, but it is found most commonly in patients with a

failing Fontan circulation [8]. The pathophysiology remains to be elucidated, but Fontan physiology is increasingly recognized as a risk factor, with onset of symptoms shortly after completion of the Fontan [6].

Multiple treatments have been proposed for the management of this condition including antibiotics, steroids, bronchodilators, mucolytic agents, anticoagulation, aerosolized urokinase, and tissue plasminogen activator, all with variable anecdotal success. Improvement of hemodynamics (e.g., via stent fenestration) and atrioventricular (A-V) pacing in the Fontan circulation have occasionally led to amelioration of this disorder [8, 10].

Case Report

A 15-year-old boy experienced intermittent bouts with shortness of breath and cough with blood and thick sputum. He had been born with hypoplastic left heart syndrome associated with mitral and aortic stenoses, which had been palliated with a nonfenestrated lateral tunnel Fontan procedure at the age of 6 years. His postoperative course included successful balloon dilation of a mild aorta recoarctation at the age of 13 years, but otherwise was unremarkable. The patient had been asymptomatic and was treated electively with lisinopril and warfarin.

A flexible fiberoptic bronchoscopy was performed, which showed a white acellular-mucinous cast, cough malacia, and moderate persistent reactive airway disease. The boy was started on a vigorous pulmonary toilet regimen including a chest physiotherapy vest, with transient improvement in pulmonary function, but with increased expectoration of large acellular casts. He was readmitted for coughing spells associated with desaturation.

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The lung examination showed markedly diminished breath sounds in the right lower lung fields. A chest x-ray showed a density in the right lower lung. Blood tests showed a normal white blood count, no anemia, and a normal platelet count. There was no growth from blood cultures produced on two occasions. Cardiac catheterization showed mean pulmonary artery pressures of 15 mmHg. Venovenous collaterals from the Fontan circuit to the left lower pulmonary vein were successfully coil embolized, with no change in pulmonary artery pressures and improvement in oxygenation.

The boy was recatheterized 3 months later because his expectoration of casts had increased. At that time, he was noted to have mean pulmonary artery pressures of 17 to 18 mmHg. Significant aorta-to-pulmonary artery collaterals were successfully coil embolized. Bronchoscopy was performed 5 days after catheterization for persistent hypoxia, at which time several large casts were removed, again with transient improvement.

With recurrence of multiple large cast expectorations 2 months later (Fig. 1), the boy was started on aerosolized tissue plasminogen activator (TPA) as an outpatient, but continued to expectorate large acellular-mucinous casts. It was decided to start carvedilol to improve cardiac function in hope of ameliorating the casts. At this writing, approximately 1 year has passed, and the boy has had only three casts. He continues to receive both cardiac and pulmonary medications without further complications.

Discussion

Plastic bronchitis has been reported in association with a variety of cardiorespiratory disorders including asthma, cystic fibrosis, allergic bronchopulmonary aspergillosis, bacterial and viral respiratory infections, sickle cell disease with acute chest syndrome, and cyanotic congenital heart disease (CHD) [1]. Cyanotic CHD is by far the most common clinical condition associated with plastic bronchitis [5]. As reported in most cases, plastic bronchitis occurs after patients undergo a vascular diversion of systemic blood flow into the pulmonary circulation as part of a staged operation to correct underlying CHD defects [5]. Casts observed after the Fontan operation often are described as acellular and composed mostly of mucin [10].

Two classification schemes have evolved, building on new evidence of cast composition and proposed etiologies. Sear et al. [10] categorized the disease into two types based on histologic makeup of the cast. Type 1 casts were identified as composed of eosinophilic and fibrinous infiltration and seen in patients with inflammatory conditions such as asthma, atopy, or bronchiolitis. Type 2, or noninflammatory, casts were identified as composed mostly of



Fig. 1 Bronchial cast

acellular-mucinous infiltration and seen primarily in cases of cyanotic CHD.

Brogan et al. [3] reclassified plastic bronchitis according to the associated disease state, proposing three categories: allergic/asthmatic, cardiac and idiopathic. Madsen et al. [5] combined the classification schemes and provided a new system based first on the associated disease and second on cast histology if comorbidity is unclear.

Onset of symptoms typically has been reported within 1 to 3 years after completion of Fontan [7]. The reported case involved a 15-year-old boy with onset of symptoms 9 years after Fontan. To our knowledge, this is the first reported case of a mucinous cast involving a cyanotic CHD with such a delay in the onset of symptoms. This raises new questions regarding the pathophysiology of acellular-mucinous plastic bronchitis. Some have proposed high pulmonary venous pressures leading to an abnormal response of respiratory epithelium and resulting in excess mucus production [10]. Others have suggested that endobronchial lymphatic leakage plays a role in cast formation [4].

Madsen et al. [5] propose two steps to cast formation. The first step involves an underlying genetic predisposition. Second, there must be an inflammatory stimulus leading to a dysregulated hypersecretion of mucin. Cardiac shunt surgery appears to serve as a trigger because most

cases of plastic bronchitis occur soon after surgery but well before long-term complications of the Fontan, such as protein-losing enteropathy [6].

Historically, no standard treatment has been recognized, although several cases have shown anecdotal success with a variety of agents including tissue plasminogen activator, urokinase, steroids, mucolytic agents, and anticoagulation [1]. Madsen et al. [5] reported three cases of children improving after atrial pacing was established [5], and although the number of reported subjects is limited, it is the most reported with successful outcomes.

Conclusion

The reported case exemplifies acellular-mucinous plastic bronchitis in a patient with Fontan physiology. We believe our reported case is the first to show such a long delay in presenting symptoms after Fontan completion and can only speculate as to why the casts did not present earlier as in most cases. The hypersecretion of mucin from bronchial epithelium and the elevated pulmonary venous pressures seem to be a common denominator in most cases of type 2 plastic bronchitis associated with Fontan physiology, but the etiology that connects the two still is not completely understood. Whether the elevated pressures lead to an abnormal response of the bronchial epithelium, resulting in excess mucus production [9] or whether there is an underlying host factor inherited or unmasked by a trigger that causes a malignant mucin hypersecretion [5], treatment should include a vigilant cardiac evaluation, with

correction of the underlying defect and optimization of cardiac output and cardiac rhythm.

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