Plastic Bronchitis

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KEYWORDS

• Mucus • Mucin • Secretory hyperresponsiveness • Cast bronchitis • Fontan physiology
• Congenital heart disease • Pulmonary lymphatics

KEY POINTS

- Plastic bronchitis associated with congenital heart disease or with lymphatic anomalies is caused by aberrant pulmonary lymphatic vessels and drainage. This true form of plastic bronchitis can usually be treated by selective lymphatic vessel ablation.
- Plastic bronchitis is probably more common than reported. This speculation is based on the observation that many clinicians are unfamiliar with the disease and may fail to recognize milder forms of the syndrome.
- Plastic bronchitis with cohesive branching airway casts should not be confused with the more common, but smaller and more etiologically distinct, casts that are associated with mucus plugging.
- For nonlymphatic plastic bronchitis associated with eosinophils and Charcot-Leyden crystals within casts, the most effective therapy seems to be cast removal followed by high-dose or pulse corticosteroids.

INTRODUCTION

Plastic bronchitis (PB) is an uncommon pulmonary disease characterized by production of cohesive and branching casts filling the airways (**Fig. 1**). This disease has been recognized for thousands of years and was first described by Galen in patients who he thought were expectorating pulmonary veins. PB has had many other names through the years, including fibrinous bronchitis and cast bronchitis.

PATIENT EVALUATION OVERVIEW

The diagnosis of PB is confirmed by a history of expectoration of branching airways casts, or by removing branching casts at the time bronchoscopy. Life-threatening respiratory distress can occur because of obstruction of airways with casts in children with congenital heart disease or as a consequence of lymphatic engorgement following surgical correction of congenital heart disease.[1] The casts of PB often contain an abundance of mucin but unlike mucin polymers in normal mucus, which are linearly linked, there is significant cross-linking between adjacent mucin strands (**Fig. 2**). Most casts have only small amounts of fibrin.[2]

True PB associated with expectoration of branching casts has not been reported in patients with cystic fibrosis or bronchiectasis. Although there are large amounts of polymeric DNA and F-actin in cystic fibrosis, this rarely, if ever, supports the formation of the complex branching casts that are diagnostic of PB.[3] Furthermore, polymeric DNA and F-actin are not abundant in PB casts, so aerosol dornase alfa is typically ineffective as therapy (discussed later). PB must also be differentiated from the mucous plugging that is associated with fungal inflammation of the airway in allergic bronchopulmonary aspergillosis.

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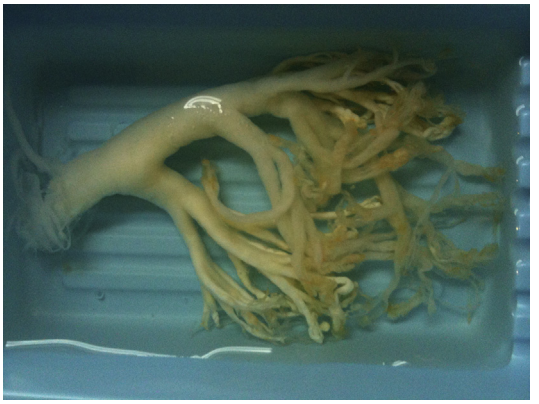

Fig. 1. Typical expectorated branching cast from a child with plastic bronchitis caused by congenital heart disease.

It is not clear whether true PB can be part of the asthma spectrum in patients with severe asthma and secretory hyperresponsiveness.[2]

The author examined PB casts from more than 50 adults and children with a variety of associated conditions. All showed the presence of inflammatory cell infiltrates that predominantly comprised lymphocytes and, at times, eosinophils. Inflammatory cells are more commonly seen in association with asthma and allergy and other non–cardiac-associated conditions.[4]

PB is underdiagnosed and may be first discovered at autopsy. Patients with milder forms of cardiogenic PB may undergo spontaneous recovery as cardiac function improves.

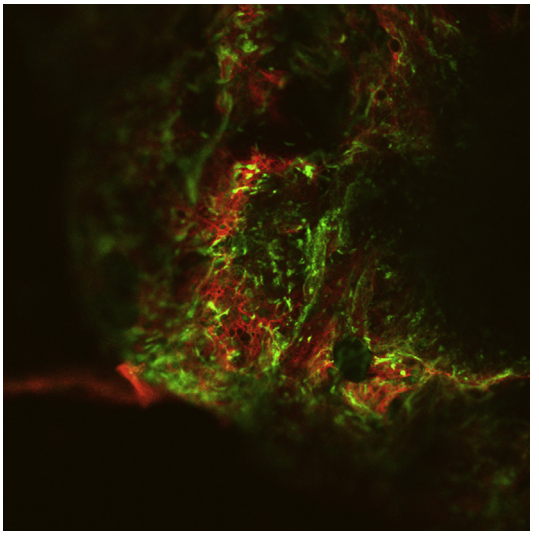

Fig. 2. Confocal microscopy showing the polymer structure of a plastic bronchitis cast. Polymeric mucin is pseudocolored red by Texas Red–UEA (Ulex europaeus agglutinin I) lectin, intracellular (nuclear) DNA is green (Yoyo-1), and fibrin polymers are blue (by immunohistochemistry).

DISEASE ASSOCIATIONS

PB in children is usually associated with congenital heart disease and especially those with single-ventricle, Fontan physiology (**Box 1**).[5] The occurrence, severity, and the frequency of exacerbations of PB vary markedly among patients with congenital heart disease, sometimes first appearing years after surgery. Some patients have subclinical disease with the expectoration of small casts or resolution of casts between exacerbations. PB has been associated with lymphatic abnormalities both in patients with congenital heart disease and in patients with primary abnormalities of lymphatic flow. MRI mapping of thoracic lymphatics has conclusively shown that all of these patients have aberrant pulmonary lymphatics and, in patients with congenital heart disease, these vessels have a distinctive appearance.[6] Ablation of these abnormal lymph vessels using direct injection of tissue glue is almost uniformly effective in reducing, and often eliminating, cast formation.[7]

A PB-like condition can be triggered by the inhalation of toxic gases, such as sulfur mustard. In experimental animals that have inhaled sulfur mustard there is extensive plugging of the airway with fibrin-rich casts that are histologically different from PB. In these animal models, casts can be effectively treated with inhaled heparin[8] or tissue plasminogen activator (tPA).[9]

Box 1
Conditions associated with plastic bronchitis

Proven conditions

Congenital heart disease with Fontan physiology

Pulmonary lymphatic anomalies

Influenza A pulmonary infection

Possible conditions

Toxic inhalation

Sickle cell acute chest syndrome

Hypersecretory and near-fatal asthma (eosinophilic casts)

Unlikely and unproven conditions

Cystic fibrosis

Chronic obstructive pulmonary disease

Bronchiectasis

Bacterial pneumonia