Plastic bronchitis: Casting a wider net.

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Abstract

As our understanding of the uncommon group of disorders called plastic bronchitis has evolved, so too has the classification of these disorders.

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As our understanding of the uncommon group of disorders called plastic bronchitis has evolved, so too has the classification of these disorders. A case series by Seear (1) subdivided these based upon cast histology as Type 1 or inflammatory casts and Type 2 containing only “mucin”. In 2005 we published a more extensive review based on underling characteristics and outcome and at that time, started a Plastic Bronchitis Registry and specimen repository; NCT 01663948 (2). Initially, nearly all subjects entered into the registry were children with congenital heart disease and single ventricle (Fontan) physiology and we showed that these casts contained neither fibrin nor mucin. Then in 2013-2014, Dori and Itkin showed that these Type 2 casts were caused by abnormal lymphatic drainage into the thoracic duct, that they were comprised of congealed lymph, and that this form of plastic bronchitis could be successfully treated by selective lymphatic embolization (3). In 2015, Joy Dumaire, an adult nurse with plastic bronchitis due to non-cardiac lymphatic abnormalities, started the Plastic Bronchitis Foundation (4) and with the presence of the Plastic Bronchitis Foundation on social media, the registry quickly expanded with many more adult patents self-reporting; although to be included in the registry the diagnosis had to be confirmed by their physician. Many of these adults had non-cardiac lymphatic dysplasia although a significant number had no lymphatic abnormalities, but rather airway casts comprised of eosinophils and their degradation products. Based on this, plastic bronchitis is now classified as either lymphatic (LPB) or eosinophilic (EoPB) (4). Plastic bronchitis is uniquely confirmed by the observation of branching airway casts and has never been shown to co-exist with CF or bronchiectasis as diseases that produce the more common sputum plugs (5)

Far less is known about EoPB and the case series published in this issue of Pediatric Pulmonology (6) both adds to our knowledge and is consistent with what we have found in the registry where 32 subjects (18 adults) have been registered with EoPB; roughly one quarter of the number reported with LPB (unpublished data). EoPB findings from the registry data include:
1. As reported in this manuscript, although some subjects have a diagnosis of asthma and allergies, many more do not, despite having eosinophils in both the airway epithelium and in the casts.

2. Few of these subjects have an increased IgE, although mild to moderate peripheral eosinophilia is not uncommon.

3. The cast formation often follows a viral LRTI.

4. In each patient, the casts have tended to form in the same airway each time, and in some patients, casting resolves spontaneously over time. Casting appears to be more often seen in the left lung although the reason for this is unknown.

5. As reported here, bronchoscopy cast extraction is best accomplished using a cryoprobe as these casts are quite friable and slippery.

6. Most adult subjects with PB (both forms) are overweight or obese.

Based on these observations, we have posited that EoPB has more in common with eosinophilic esophagitis than with asthma. We have had reasonably good therapeutic success using pulse steroid therapy to reduce casing, and no success at all using mucolytics. There have now been case reports of successful treatment of EoPB with mepolizumab (7) and it is likely that this, or another biologic effective in treating tissue eosinophilia, will become the preferred therapy for this disorder.

References


[6] Current paper this issue