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Case Report

Asthma-Related Plastic Bronchitis: A Challenging Diagnosis

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Abstract

Plastic bronchitis (PB) is a rare but potentially life-threatening condition characterized by the formation of bronchial casts, leading to airway obstruction. It is associated with several underlying conditions, including congenital heart disease, respiratory infections, and asthma. Due to its non-specific symptoms, PB is often misdiagnosed or underdiagnosed. We present a case of a 10-year-old boy with a one-year history of recurrent productive cough, dyspnea, and fever, unresponsive to albuterol treatment. Chest X-rays revealed atelectasis of the left lower lung, which persisted despite airway clearance techniques. High-resolution computed tomography (HRCT) showed collapse consolidation of the left lung. Bronchoscopy revealed bronchial obstruction by a rubbery cast, which was successfully removed. Pathologic analysis of the cast showed eosinophils, neutrophils, and Charcot-Leyden crystals. The patient was treated with inhaled corticosteroids and short-acting beta-agonists, leading to complete resolution of symptoms. Follow-up after six months showed no recurrence of symptoms. This case highlights the importance of considering PB in patients with recurrent respiratory symptoms and atelectasis, particularly when foreign body aspiration or common respiratory diseases have been ruled out. Early diagnosis and management, including cast removal and treatment of underlying conditions, are crucial to preventing complications.

Keywords: Plastic Bronchitis; Infections; Asthma; Respiratory Diseases

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Introduction

Plastic bronchitis (PB) is a severe though uncommon pulmonary condition characterized by the presence of cast in branching intrabronchial tree and subsequent blockage of distal airways (1). The underlying mechanism of cast formation in PB is unknown. However, associations of PB with surgical corrections of congenital heart diseases and other cardiac procedures like Fontan and Glenn shunt (hemi-Fontan) procedures, sickle cell disease, bacterial and viral infections, inflammatory processes such as asthma and idiopathic chronic eosinophilic pneumonia and cystic fibrosis (CF) in the pediatric population as well as infections, cardiopulmonary procedures, anatomic variations in lymphatic flow, medications and environmental exposures in adults have been reported (1-8). The exact prevalence of PB is unknown due to the unusual nature of it and it seems that many patients are not diagnosed (9).

PB affects patients of all age groups with a predominance in children (9).

Patients with PB present with nonspecific symptoms including productive cough, dyspnea, wheezing, fever, or pleuritic chest pain, decreased breath sounds in physical examination, and bronchial obstruction with possible lobar atelectasis in chest radiographs (1). Differentiation between PB and other chronic respiratory illnesses like asthma, CF, or bronchiectasis could be made through the presence of branching casts in bronchoscopic evaluation (9). Acute therapy for PB includes the removal or facilitated expectoration of casts with specific treatments addressing the underlying hypersecretory process (10, 11). Removal of casts could be made mechanically through bronchoscopy; physical therapy or high-frequency chest wall oscillation. Furthermore, inhaled treatment using bronchodilators, corticosteroids, or mucolytics can disrupt the bronchial cast formation (10, 11).

Herein, we report a pediatric patient with a one-year history of productive cough and dyspnea who had atelectasis of the left lower lung in a chest X-ray. After initial evaluations to rule out the differential diagnoses, he was found to have PB, which was successfully managed through cast removal by bronchoscopy, inhaled corticosteroids, and short-acting beta-agonists.

Case presentation

A 10-year-old boy was admitted to our hospital due to a productive cough, dyspnea, and fever. He had a history of five episodes of dyspnea from one year before our visit, each episode lasted for 6-8 weeks. He was administered with albuterol by his primary care physician; the dyspnea improved partially, but not completely, using the albuterol. His mother has a history of allergic rhinitis and his father and brother were positive for asthma and were under treatment with albuterol. His recent episode of dyspnea was initiated 3 months before, he was admitted due to the addition of fever and productive cough to the dyspnea. On the day of admission, he had a fever (T=39.1), shortness of breath, with a respiratory rate of 17 and O2 saturation of 96%, and decreased breath sounds of the left lower lobe. The patient's weight and height were 30 kilograms and 137 centimeters. His chest X-ray (CXR) obtained on his admission, one month and two months before admission revealed atelectasis of the left lower lung **(Figure 1A, B,** and **C)**. His laboratory tests revealed an increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Other than the eosinophil level of 16000, the patient's complete blood count (CBC) parameters were within normal range. He was administered intravenous antibiotics with a probable diagnosis of lower respiratory tract infection. The fever was resolved and the productive cough and dyspnea improved following the antibiotic therapy. Despite performing an airway clearance technique, the left lower lobe atelectasis was not resolved. Consequently, high-resolution computed tomography (HRCT) was obtained which showed collapse consolidation of the left lower lung **(Figure 2)**.

He was a candidate for bronchoscopy for evaluating anatomical variations resulting in obstruction, ruling out infectious diseases, and the presence of mucus plugs. He underwent fibro optic and rigid bronchoscopy that showed left bronchi obstruction by a yellowish, sticky, and rubbery material which was removed **(Figure 3A** and **3B)**. Following the cast bronchitis removal, The CXR and HRCT showed resolved atelectasis (Figure 1D). The pathologic evaluations of the cast showed numerous eosinophils, neutrophils, and Charcot-Leyden crystals in a fibrotic background.

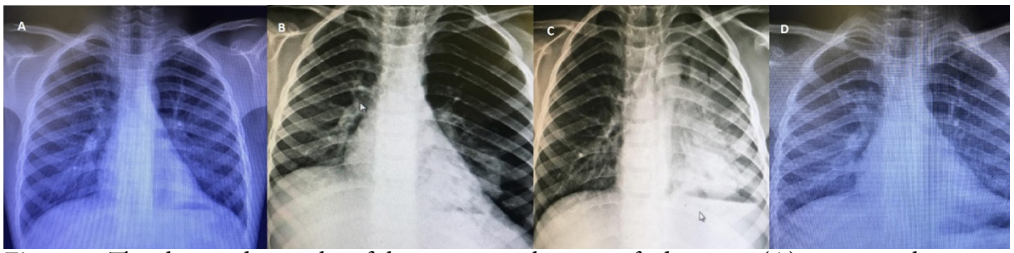

Figure 1. The chest radiography of the patient at the time of admission (A), one month preior to admisssion (B) and two months prior to admission (C) showing the atelectasis of the left lower lung and the resolved atelectasis follwing the cast removal by bronchoscopy (D).

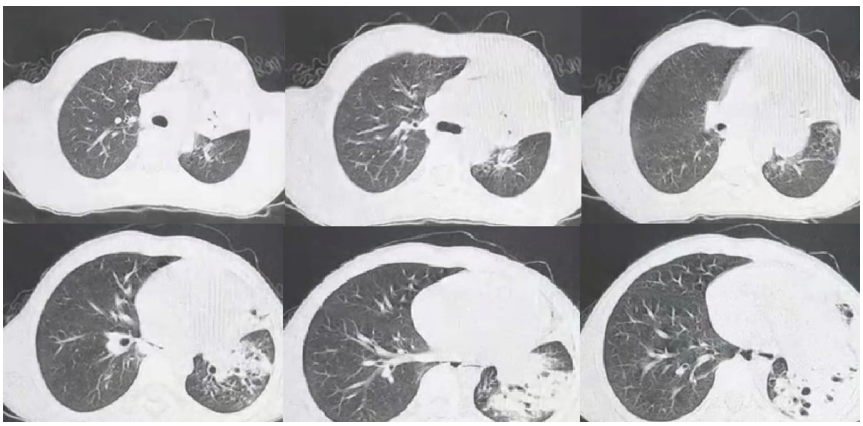

Figure 2. The High Resolution Computed Tomography (HRCT) of the patient before cast removal, indicating atelectasis of the left lower lung

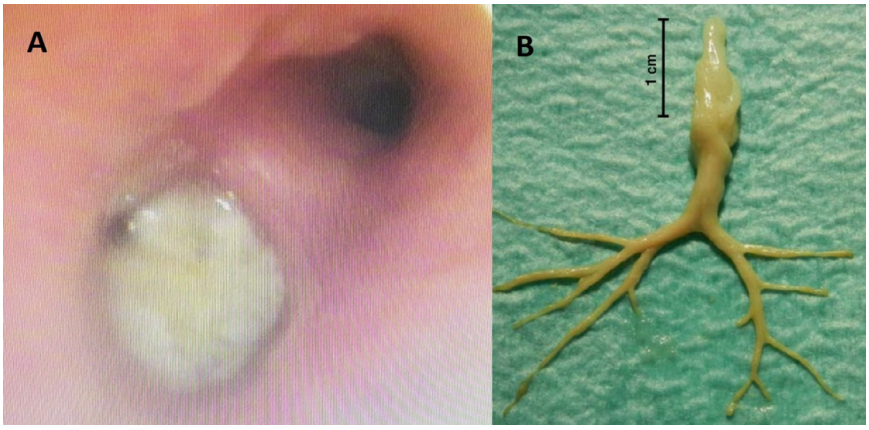

Figure 3. The bronchoscopic view of the cast (A) and the removed cast with the structural bronchial tree (B).

Differential diagnosis

Tuberculosis (TB)

It has been estimated that 5%-10% of children with latent TB infection, become symptomatic, with lung involvement as the most common site of infection (12). Common physical examinations in these patients consist of fever, weight loss, night sweats, cough, and hemoptysis. Hilar or mediastinal lymphadenopathy in these patients could cause significant mass effects on the adjacent pulmonary trees, resulting in the post-stenotic collapse-consolidation patterns seen in CXR. The described patient had a history of a one-year cough besides the atelectasis of the left lower lung, with no history of malaise, weight loss, night sweats, or hemoptysis. The purified protein derivative (PPD) test and Broncho alveolar lavage (BAL) were performed to rule out TB and Acid-fast bacillus (AFB), which were negative.

Cystic Fibrosis (CF)

CF is a multisystem disorder, affecting the respiratory system, gastrointestinal (GI) reproductive tract, and sweat glands. Although it is a multisystem disease, the respiratory tract and lungs are mostly involved. Failure to thrive (FTT), chronic cough, and wheezing are among the most common clinical presentations of CF in children (13). The mucus plugging in these patients could lead to atelectasis. The common findings in HRCT and CXR include air-trapping, bronchiectasis, mucus plugging, and bronchial dilation. The described patient did not have FTT or wheezing and the only radiologic finding in favor of CF was atelectasis due to mucus plug. The CF was ruled out for the patient by a negative sweat chloride test.

Primary ciliary dyskinesia (PCD)

PCD is an autosomal recessive disorder of mucociliary function. Incompetent mucociliary clearance, mucus retention, and consequent mucus plug formation occur as a result of PCD (14). The main clinical presentations of PCD include chronic respiratory infection, infertility, and situs

inversus (14). Patients often complain of productive cough and nose congestion (15). Its diagnosis is based on the combination of the clinical features and nasal nitric oxide (NO) level. Besides, a questionnaire-based prediction tool (PICADOR) has been developed for prediction of the risk of the presence of PCD (16). Both the nasal NO level and PICADAR score in the presented patient were not in favor of the presence of the PCD.

Allergic bronchopulmonary aspergillosis (ABPA)

ABPA is considered a pulmonary disorder caused by Aspergillus with not clearly understood pathophysiology. Its clinical manifestations consist of fever, weight loss, poorly controlled asthma, increased productive cough increment, and dyspnea (17). Mucus plugs are the main radiological findings in these patients (17). The diagnosis is based on the clinical findings, serum total immunoglobulin E (IgE) level, and cutaneous reactivity to Aspergillus fumigatus. As the findings of the described patient were highly suggestive of ABPA, serum total IgE level and cutaneous reactivity to Aspergillus fumigatus were evaluated and both were negative.

Bronchiectasis

Bronchiectasis unrelated to CF causes irreversible bronchial dilatation characterized by chronic productive cough, recurrent pneumonia, dyspnea, clubbing, and coarse crackles (18). Nearly 40% of bronchiectasis occurs with no specific underlying etiology (19). The Gold standard for diagnosing bronchiectasis is lung HRCT, showing multilobar involvement as bronchial dilation, increased broncho-arterial ratio, bronchial wall thickening, and lack of bronchial tapering (20). The described patient had a history of prolonged dry cough and dyspnea, without recurrent pneumonia and clubbing. He did not have coarse crackle on lung auscultation and on HRCT, no bronchial dilation, and increased broncho-arterial ratio and bronchial wall thickening were noted; therefore, Bronchiectasis unrelated to CF was not a possible diagnosis.

Foreign Body Aspiration

Another probable diagnosis could be foreign body aspiration, leading to left lower lobe atelectasis; with possible superimposition of pneumonia. The common clinical presentations of foreign body aspiration include recurrent cough or wheezing, fever, bronchiolitis, and asthma (21). It is more prevalent among children with less than 3 years of age (21). The diagnostic and treatment gold standard is foreign body removal by rigid bronchoscopy. The clinical symptoms of the described patient were in line with foreign-body aspiration; however, the patient was 10 years old and did not remember any history of foreign-body aspiration and choking. A rigid bronchoscopy was performed to rule out this diagnosis, which showed no foreign body.

Plastic Bronchitis (PB)

PB is a severe respiratory disorder, characterized by mucofibrinous bronchial cast formation resulting in partial or complete bronchial lumen obstruction (22). Clinical manifestations vary from mild symptoms to severe life-threatening ones leading to death. Similar to foreign body aspiration, the usual presentations include productive cough, dyspnea, fever, wheezing, respiratory distress, or chest pain and in the worst-case scenario, it causes airway obstruction, asphyxiation, and even sudden death (23). The radiologic studies show consolidation and atelectasis secondary to airway obstruction and contralateral lung hyperinflation (24). The diagnostic hallmark includes the presence of casts in the bronchial tree identified and removed by bronchoscopy (24). The clinical manifestations of the presented patient were productive cough, fever, progressive dyspnea with partial response to albuterol, and decreased lung sound on auscultation. Furthermore, radiologic findings included atelectasis, and bronchoscopy findings revealed cast. All evidence was in line with the diagnosis of PB.

Management of the patient

Following the cast removal by bronchoscopy, the patient was discharged with Inhaled corticosteroids and short-acting beta-agonists. The CXR and HRCT studies found that the atelectasis and consolidation of the left lower lung were resolved after bronchoscopic cast removal. On the 6-month follow-up visit, the asthma was controlled with the prescribed treatment with no history of fever, dyspnea, shortness of breath, or cough.

Discussion

Both adults and children could be affected by PB, but most described cases in the literature are children (11, 24). Its prevalence of PB is unknown due to its non-specific signs and symptoms and difficult-to-diagnose nature. It could be a complication of some medical conditions, such as asthma, cystic fibrosis, sickle cell anemia, lymphatic abnormalities, infections, and congenital heart surgery (Fontan procedure) (24). PB was associated with the history of Fontan procedure in most patients; its association with asthma is rare. One recent study in the pediatric population with PB showed that 70.5% of the patients had a history of Fontan procedure, 26.5% had previous respiratory diseases and 3% had idiopathic PB; the prevalence of asthma-related PB was 17.6% in their study (24). Based on the previous studies, 28%-60% of cardiac-related PB and 6%-7% of non-cardiac-related PB died as a result of this condition (25); therefore, its prompt diagnosis and management are crucial.

The goal of treatment in these patients is cast removal, symptom alleviation, and future cast formation prevention. Given different PB etiologies, the treatment options vary remarkably according to patient-to-patient basis. The overall treatment includes the combination of pharmacologic (including administration of corticosteroids, acetylcysteine, heparin, and tissue plasminogen), nonpharmacologic treatment (such as chest physiotherapy), and bronchoscopic procedures (11, 20, 22, 24). Chest physiotherapy and cast removal by bronchoscopy are the most common initial therapies; the definite treatment that may prevent future cast formation is treating the underlying conditions. Most of the patients with asthma-related PB improve following cast removal and administration of inhaled or systemic corticosteroids (24). The positive effects of corticosteroids in these patients could be attributable to both their effect on asthma control and their anti-inflammatory characteristics which prevent eosinophils infiltration (26). Our patient was administered with inhaled corticosteroid and albuterol following the cast removal. The asthma was controlled with these agents and the patient's follow-up showed no recurrence of the casts which is consistent with favorable effects of corticosteroid consumption.

Conclusion

PB is a rare entity but should be considered in patients with recurrent episodes of dyspnea and dry cough unresponsive to the treatment and those for whom foreign body aspiration is suspected. Of Note, if the previous history of Fontan procedure or other inflammatory respiratory diseases such as asthma exists, the diagnosis of PB becomes more probable.

Conflict of interests

There is no conflict of interest.

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