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Clinical and Radiological Evaluation and Follow-Up of Patients with Noncardiac Plastic Bronchitis

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What is already known on this topic?

- *Plastic bronchitis is a rare disease characterized by casts filling the airways. The pathophysiology of the disease is unknown. Cardiac surgery (Fontan or Blalock–Taussig operation), cyanotic congenital heart disease, asthma, allergic bronchopulmonary aspergillosis, cystic fibrosis, bronchiectasis, bacterial pneumonia, and sickle cell anemia are the most common etiologies.*

What this study adds on this topic?

- *We described 6 patients with noncardiac plastic bronchitis. Plastic bronchitis is very rare in children. This study adds clinical–radiological features and long-term outcomes of children with plastic bronchitis on this topic.*

ABSTRACT

Objective: Plastic bronchitis (PB) is a rare disease characterized by obstruction of the airway by fibrinous mucus plugs. The etiology can be idiopathic or secondary to systematic diseases such as congenital heart diseases. Definitive diagnosis is made by pathological examination of the sputum or bronchial sample taken by bronchoscopy. In this study, the clinical status and treatment status of patients with PB were evaluated.

Materials and Methods: Medical records of the patients diagnosed as PB were reviewed retrospectively. Age, gender, clinical symptoms, radiology, bronchoscopic findings, and pathology results were documented.

Results: Six patients with PB were included in this study (female:male, 2:4). The median age of the diagnosis was 45 months. The most common symptoms are persistent wet cough and shortness of breath. The duration of symptoms ranged from 30 to 90 days. Atelectasis was the most common radiological finding. Diagnosis was made with pathological examination of the mucus in all patients. All of the patients were treated with bronchoscopic removal of the mucus, and 4 patients required oral prednisolone therapy. Symptoms and radiological findings resolved completely in all patients.

Conclusion: Although PB is a rare disease, it should be kept in mind in relation to patients with persistent radiological and clinical respiratory symptoms.

Keywords: Plastic bronchitis, bronchoscopy, chronic cough, atelectasis, mucus plugs, cast

INTRODUCTION

Plastic bronchitis (PB) is an uncommon disease characterized by formation of fibrinous mucus plugs.[1] Fibrinous bronchitis, pseudomembranous bronchitis, and Hoffman's bronchitis are historical names of the disease.[2] A treelike bronchial cast formation is the characteristic appearance in PB. Mucus plugs can be classified into 2 types: type 1 or inflammatory casts are characterized by a dense inflammatory infiltrate composed of fibrin and eosinophils and type 2 or noninflammatory casts are found in patients with congenital heart diseases. The pathophysiology of the disease is unknown. Most of the cases are idiopathic. Cardiac surgery (Fontan or Blalock–Taussig operation), cyanotic congenital heart disease, asthma, allergic bronchopulmonary aspergillosis, cystic fibrosis, bronchiectasis, bacterial pneumonia, and sickle cell anemia are the most common etiologies.[3,4] The most common symptoms are persistent wet cough and shortness of breath.[5] In physical examination, wheezing or

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Table 1. Demographic and Clinical Findings of Patients with Plastic Bronchitis

	Patient 1	**Patient 2**	**Patient 3**	**Patient 4**	**Patient 5**	**Patient 6**
Age (months)	48	54	96	84	54	13
Diagnosed age (months)	36	18	72	84	54	13
Gender	Male	Female	Female	Male	Male	Male
Any chronic disease	No	Asthma	No	No	No	No
Symptom	Wet cough	Wet cough	Shortness of breath	Wet cough	Shortness of breath, wet cough	Wet cough
Duration of symptoms (days)	30	90	30	90	60	30
History of respiratory problems	Yes	Yes	Yes	No	No	No
History of asthma in family	No	Yes	No	No	No	No
Atopy in family	Yes	No	No	No	No	No
Oxygen saturation (%)	95	96	96	94	97	98
X- ray	Pneumothorax, atelectasis at left upper zone	Aeration differences, atelectasis at left lower zone	Atelectasis at left upper and lower zone	Total atelectasis at left side	Opacity at right middle zone	Opacity at left upper zone
Computed tomography	Pneumothorax atelectasis at left upper lobe	Ground-glass opacity peribronchial thickening, atelectasis at left lower lobe	Not performed	Total atelectasis at left upper and lower lobes	Consolidation, atelectasis, and mucus plugs at right upper lobe	Atelectasis at upper left lobe and air bronchogram
Findings of bronchoscopy	Mucus plugs at left upper lobe and basal segments	Bronch-shaped cast and occlusive mucus plugs at left lower lobe	Occlusive mucus plugs at left upper and lower lobes	Occlusive mucus plugs along the left main bronchus	Mucus plug and bronchioleshaped cast in right upper	Mucus plug and treelike bronchial cast at left upper lobe entrance
Treatment	Methylprednisolone and inhaled corticosteroid (6 months)	Methylprednisolone (6 months) ICS (on treatment)	Methylprednisolone (6 months)	Methylprednisolone (6 months)	No drug treatment*	No drug treatment*
Time after treatment (months)	12	42	26	12	24	36
Recurrence	No	Yes**	No	No	No	No

ICS, inhaled corticosteroid.
*Total recovery after removal of local mucus plugs.**Total recovery after 2-week steroid treatment.

decreased breath sounds can be heard. Some patients may be misdiagnosed as foreign body aspiration or asthma.[6]

Chest x-ray is the first tool for diagnosis. Atelectasis and persistent infiltrations can be seen on chest x-ray.[7] Computerized tomography (CT) can be performed for a differential diagnosis.

A definitive diagnosis is made by pathological examination of the sputum or bronchial sample taken by bronchoscopy. The first step of the treatment is the removal of mucus plugs with bronchoscopy.[8] Sometimes, mucus plugs can be expectorated by patients spontaneously. Corticosteroids, antibiotics, and inhaled treatment can be used for treatment. In this study, we aimed to demonstrate the clinical features and long-term follow-up results of patients with PB.

MATERIALS AND METHODS

All patients diagnosed with PB at our Pediatric Pulmonology Department between 2012 and 2019 were included in this study. The medical records of the patients with PB were reviewed retrospectively. Age (median, minimum, and maximum), gender, clinical symptoms, radiological and bronchoscopic findings, pathology, and treatment results were evaluated in all cases. Flexible bronchoscopy (FB) was performed on all patients by an experienced pediatric pulmonologist. Flexible bronchoscopy was performed in the operating room. Sedation and anesthesia were administered to all patients by an anesthesiologist. Midazolam was used for premedication, and propofol was used for sedation and anesthesia. A 3.6 mm diameter Fujinon bronchoscope was used in all operations. Lidocaine was used as topical anesthesia for the vocal cords and carina. In all patients, the nasal route was used. During the procedure, oxygen supplementation was provided for all patients. Bronchoalveolar lavage (BAL) specimens of patients were evaluated microbiologically and pathologically. Pretreatment and posttreatment radiological examination were evaluated by same experienced radiologist.

This study was approved by the Bezmialem Vakif University Ethics Committee (July 7, 2020, approval no. 11/237).

RESULTS

Six patients with PB were included in this study (female:male, 2:4). None of the patients had congenital heart disease. One patient had a family member with asthma. The median age of the patients at diagnosis was 45 months (minimum: 13 months; maximum: 84 months). Five patients had chronic wet cough, and 2 patients had shortness of breath. The duration of symptoms ranged from 30 to 90 days (median: 55 days). Only 1 patient had asthma (Table 1).

Atelectasis was the most common radiological finding (%100) in radiology (Figures 1A and 1B). Consolidation, pneumothorax, and aeration differences were also seen. All patients underwent flexible bronchoscopy by an experienced pediatric pulmonologist. There was no major complication during the bronchoscopy procedure. Five patients had a treelike bronchial cast plugs on the left side (Table 1) (Figures 1C and 1D). Fibrin-rich necrotic tissue, mucinous material, and inflammatory and eosinophil cells, compatible with PB, were found in the pathological examinations of all patients. Two of the patients had no additional treatment after the total removal of mucus plugs with bronchoscopy. Four patients with bilateral and severe involvement who had severe, recurrent clinical symptoms and eosinophilia in their BAL specimen were treated with methylprednisolone for 6 months. Methylprednisolone was started at a dose of 2 mg/kg, and the treatment was continued by reducing the dose after 1 month. These patients continued on inhaled steroids for 6 months. Symptoms and radiological findings resolved in all patients.

DISCUSSION

PB is a very rare disease that is more frequently seen in children than adults. The prevalence is unknown in the pediatric population. Symptoms as wet cough, shortness of breath, and wheezing are caused by obstruction of the airway with bronchiole-shaped mucus. Congenital heart disease and lymphatic surgery are associated with PB. One study including 34 patients with PB showed that 70% of patients had a history of congenital heart disease, 9 of the patients had respiratory disorders, and 1 patient did not have an underlying disease.[9] A study that

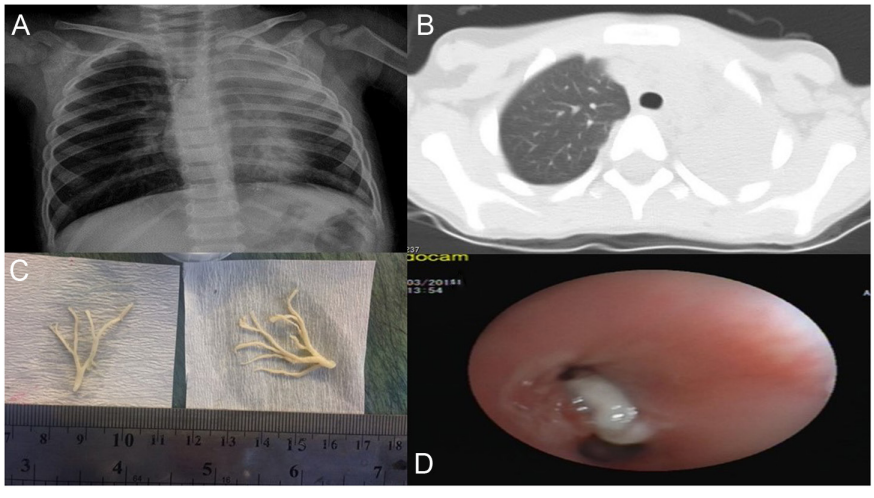

Figure 1. (A-B) Atelectasis on left upper lobe. (C-D) Bronch-shaped cast.

evaluated patients with PB without structural heart disease showed that most of their patients had asthma diagnoses and asthma is a risk factor for recurrent PB. In our study, 5 patients did not have any underlying disease, except for 1 patient with asthma who has recurrence history.[10]

Persistent wheezing, chronic wet cough, or life-threatening respiratory failure are the most common symptoms of the patients.[1,2] Chronic wet cough is also the most common symptom of our patients. The duration of symptoms can vary from patient to patient. The duration of symptoms in our patients ranged from 30 days to 90 days. Half of the patients had a history of inhaled treatment.

Atelectasis is one of the most common radiological findings in PB. Infiltration like pneumonia, ground-glass opacity, and increased aeration can also be seen.[11] Computed tomography is performed in most patients for differential diagnosis. Atelectasis and opacity can be seen on a CT scan. Chest x-ray and CT findings are mostly nonspecific and are not diagnostic alone.[9] All patients in our study had atelectasis; 2 patients had opacity, and 1 patient had pneumothorax. Most of our patients had left lung involvement. A study from China showed that 46.5% of their patients with PB had right lung involvement and 14% had bilateral involvement.[12]

Definitive diagnosis is made by pathological examination of the sample removed by bronchoscopy.[5] A treelike bronchial cast is mostly seen in bronchoscopy. Necrotic, necrobiotic fibrinous, mucinous material, and inflammatory cells are detected on pathological examination.[3] Bronchoscopy is also a treatment tool for the removal of the cast. All of our patients underwent flexible bronchoscopy. A treelike bronchial cast was observed and removed with bronchoscopy.

There is no standard medical treatment for PB. Inhaled dornase alfa, inhaled tissue plasminogen activation, inhaled *N*-acetylcysteine, and inhaled hypertonic saline, especially in patients with cardiac diseases, can be used in the treatment of PB.[9,13-15] Inhaled or systemic corticosteroids were shown to be useful in the treatment of PB.[4,9] Two patients in our series have been treated with inhaled and systemic corticosteroids. Two patients used only oral corticosteroids and 2 patients had no treatment.

The current study had some limitations: This study is a retrospective study and had a small group of patients.

CONCLUSION

In conclusion, PB is a rare disease associated with increased morbidity and should be considered in patients with chronic respiratory symptoms and persistent radiological findings.

Ethics Committee Approval: This study was approved by Ethics Committee of Bezmialem Vakif University, (Approval No: 11/237, Date: July 7, 2020).

Informed Consent: Written informed consent was obtained from the patients who agreed to take part in the study.

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