

Fatal plastic bronchitis with eosinophilic casts in a previously healthy child

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= Abstract =

Plastic bronchitis is a rare disease characterized by the recurrent formation of branching mucoid bronchial casts that are large and more cohesive than those that occur in ordinary mucus plugging. Casts may vary in size and can be spontaneously expectorated, but some require bronchoscopy for removal. Plastic bronchitis can therefore present as an acute life-threatening emergency if obstruction of the major airways occurs. Three of 22 reported patients with eosinophilic casts were fatal, with death due to central airway obstruction. Here, we report a child with no history of atopy, allergy, or congenital heart disease who was diagnosed with plastic bronchitis with eosinophilic casts. Although he was administered intravenous (iv) antibiotics; iv corticosteroids; and a vigorous pulmonary toilet regimen, including chest physiotherapy and routine bronchoscopic removal of casts, he had brain death secondary to hypoxic brain damage. Plastic bronchitis can be fatal when casts obstruct the major airways, as in the present case. Clinicians should intervene early if a patient exhibits signs and symptoms consistent with plastic bronchitis. (*Korean J Pediatr* 2009;52:1048-1052)

Key Words : Plastic bronchitis, Cast bronchitis, Airway obstruction, Eosinophilic cast

Introduction

Plastic bronchitis is a rare disease characterized by the recurrent formation of branching, mucoid bronchial casts¹⁾. Plastic bronchitis can present as an acute, life-threatening emergency if obstruction of the major airways occurs²⁾. Type I or inflammatory casts consist predominantly of fibrin with dense eosinophilic infiltrates and Type II or acellular casts are composed primarily of mucin³⁾. The prognosis is favorable in the group of patients with eosinophilic casts, but 3 of 22 reported patients with eosinophilic casts died due to central airway obstruction¹⁾. Herein, we report a child with no history of atopy, allergy, or congenital heart disease (CHD) who was diagnosed with plastic bronchitis with eosinophilic casts. Although he was given intravenous (iv) antibiotics, iv corticosteroids, a

vigorous pulmonary toilet regimen, including chest physiotherapy and routine bronchoscopic removal of casts, he had brain death secondary to hypoxic brain damage.

Case report

An 11-year-old boy was admitted to Chonnam National University Hospital for evaluation of cough of 2 days duration. His parents had previously taken him to a primary care physician. The physician evaluated the plain radiographs, which revealed total atelectasis of the left lung. The patient had no history suggestive of allergic diseases, such as wheezing, chronic cough, rhinorrhea, or atopic dermatitis and he had no admission history. On physical examination, his weight was 34.5 kg (25–50 percentiles) and he was 142 cm in height (25–50 percentiles). His blood pressure was 100/60 mmHg (25–50 percentiles), the pulse rate was 78/min, the respiratory rate was 20/min, and the body temperature was 36.5°C. He had no signs of respiratory difficulty, such as chest retractions, dyspnea, and nasal flaring. On auscultation of the chest, breath sounds were not heard over the left lung field, however clear breath

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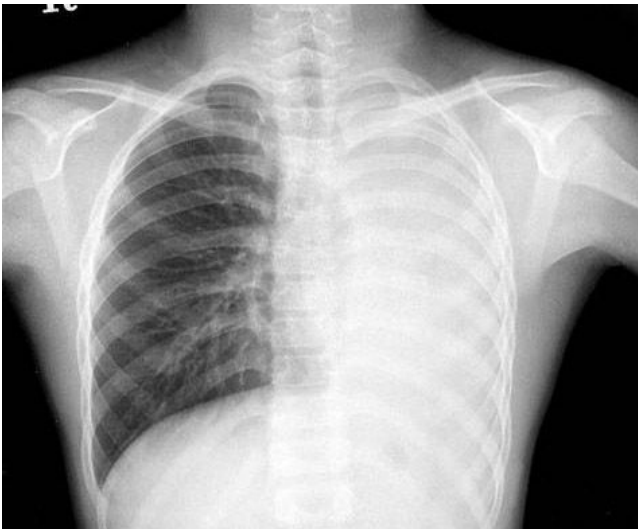


Fig. 1. On the day of admission, a thoracic roentgenogram showed total atelectasis of the left lung, overinflation of the right lung, and an abruptly truncated left main bronchus.

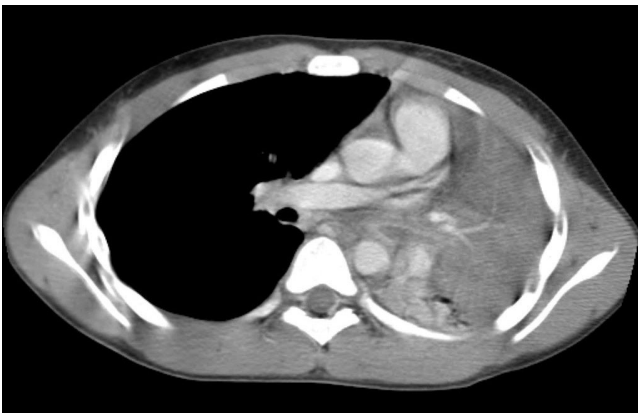


Fig. 2. On the day of admission, a chest CT showed left main bronchial obstruction with atelectasis of the left lung and compensatory overinflation of the right lung.

sounds were heard over the right lung field. Laboratory studies revealed a hemoglobin of 14.5 g/dL, a white blood cell count of $11,400/\text{mm}^3$ (polymorphonuclear cells, 70%; lymphocytes, 17%; and eosinophils, 5%), a platelet count of $424,000/\text{mm}^3$, the C-reactive protein level was 0.9 mg/dL, the erythrocyte sedimentation rate was 22 mm/hr, the lactate dehydrogenase (LDH) was 550 U/L, the anti-streptolysin O titer was 230 IU/mL, and the anti-*Mycoplasma* antibody titer was 1:40. Electrolytes, and liver and kidney function tests were in the normal range.

A chest radiograph showed total atelectasis of the left lung, overinflation of the right lung, and an abruptly truncated left main bronchus (Fig. 1). A chest CT scan showed left main bronchial obstruction with atelectasis of the left lung with an ipsilateral small pleural effusion and compensatory overinflation of the right lung (Fig. 2). An electrocardiogram and echocardiography were normal. He underwent thoracentesis. The pleural fluid was turbid and bright yellow in color. The results of the pleural fluid studies were as follows: pH, 7.38; white blood cell count, $2,200/\text{mm}^3$ with 25% neutrophils and 20% eosinophils; total protein, 4.7 g/dL; glucose, 90 mg/dL; LDH, 447 U/L; and adenosine deaminase, 14.0 IU/L. Gram-stain, acid fast stain, and potassium hydroxide (KOH) mounts of the pleural fluid were negative. Polymerase chain reaction and bacteriologic culture for *Mycobacterium tuberculosis*, and bacterial culture of the pleural fluid were negative; serial *M. tuberculosis* and bacterial cultures of the blood were negative. Shell vial culture for influenzae A and B, parainfluenzae 1, 2, and 3, adenovirus, and respiratory syncytial virus were negative. Mutational analysis of the CFTR gene

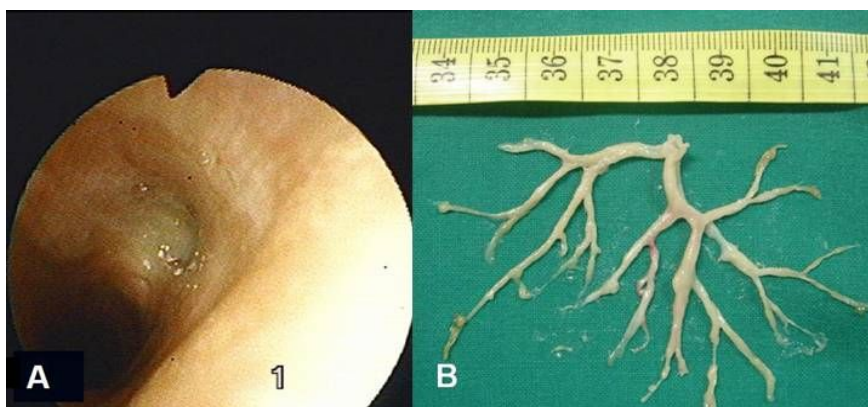


Fig. 3. (A) An emergency flexible bronchoscopy showed obstructive secretions in the left main bronchus. (B) Multiple extracted bronchial casts had a firm, rubbery consistency, and anatomy of the bronchial tree was preserved by routine bronchoscopic removal.

for cystic fibrosis was normal. An emergency flexible bronchoscopy showed obstructive secretions in the left main bronchus (Fig. 3A). A 3rd generation cephalosporin and clindamycin were administered iv, along with oral roxithromycin, and vigorous pulmonary toilet regimen, including a chest physiotherapy, was begun. He had a sudden deterioration on the 3rd hospital day. He was noted to have the acute onset of tachypnea, inspiratory stridor, decreased breath sounds in the right lung field, and wheezing in the left lung field. On physical examination, his SpO₂ measurements fluctuated between 85 and 95% on 100% face mask oxygen, and the arterial blood gas analysis was as follows: pH, 7.13; pCO₂, 109 mmHg; and pO₂, 45 mmHg. His temperature was 38.3°C, the pulse rate was 139/min, the respiratory rate was 40/min, and the blood pressure was 160/90 mmHg. Under general anesthesia, emergency rigid bronchoscopy was performed, which revealed complete occlusion of the right main bronchus by a large white adherent mucus plug, characteristic of plastic bronchitis (Fig. 3B). On pathologic examination, the firm and rubbery casts were composed of mucinous and fibrinous material containing diffuse infiltrated eosinophils (Fig. 4). After bronchoscopic removal of the cast, the dyspnea improved; however, he had intermittent dyspnea attacks and hypoxemia. As a result, he was intubated and a tracheostomy was performed. Daily routine bronchoscopy

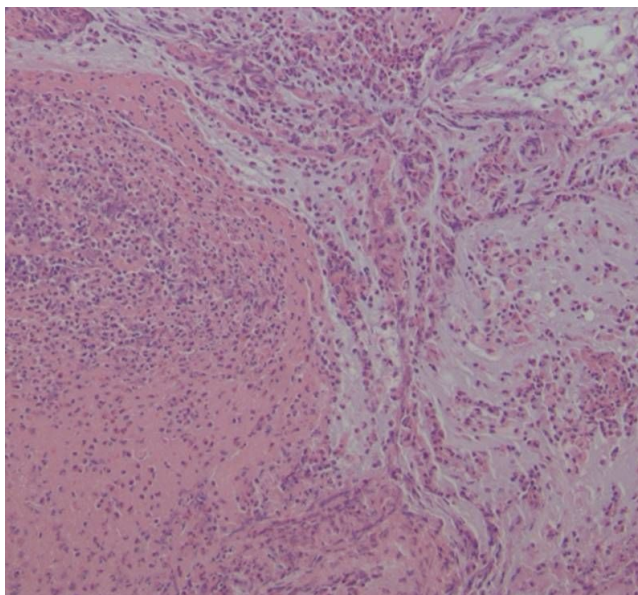


Fig. 4. Hematoxylin and eosin-stained sections (original magnification $\times 100$) showed mucinous and fibrinous material containing diffuse infiltrated eosinophils.

was performed. He continued to expectorate bronchial casts, despite iv antibiotics, iv corticosteroids, and iv mucolytics until the 6th hospital day. A follow-up chest CT showed near complete improvement of the atelectasis of the left lung, but segmental atelectasis in the right upper lung (Fig. 5). Although his lung function improved after routine bronchoscopic cast removal, he had brain death secondary to hypoxic brain damage after the 7th hospital day and died on the 41st hospital day.

Discussion

Plastic bronchitis has been referred to by many names, including fibrinous bronchitis, cast bronchitis, bronchitis pseudomembranosa, and Hoffmann bronchitis²⁾. Plastic bronchitis in children has been reported in association with a variety of cardiorespiratory disorders, such as asthma, cystic fibrosis, allergic bronchopulmonary aspergillosis, bacterial and viral respiratory infections, acute chest syndrome associated with sickle cell disease, and CHD⁴⁾. The prevalence of plastic bronchitis is unknown, and it is likely that many patients go undiagnosed. Plastic bronchitis affects patients of all age groups, but the majority of published reports involve pediatric patients²⁾.

The clinical hallmark of plastic bronchitis is the frequent formation and expectoration of very large and branching bronchial casts. These may vary in size from small segmental casts of a bronchus, to casts filling the airways of an entire lung. It has also been reported that expectorated

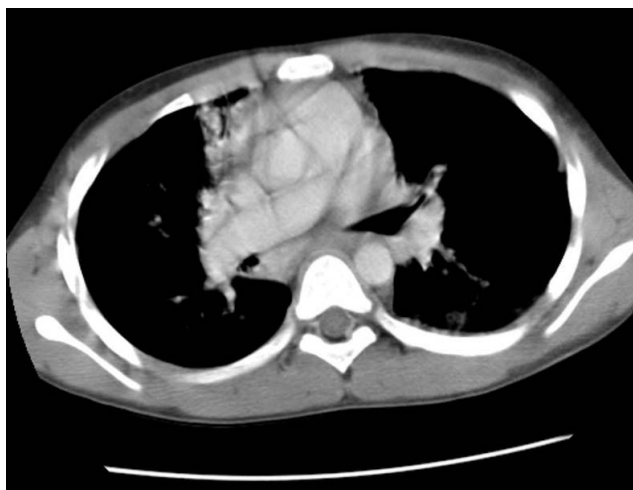


Fig. 5. A follow-up chest CT showed near-complete improvement of atelectasis of the left lung but segmental atelectasis in the right upper lung after bronchoscopic removal of the casts.

casts have been mistaken for food, such as noodles or chicken meat⁵. The bronchial casts are often described as 'pudding-like' or 'toothpaste' and more cohesive and of rubbery consistency than those which characterize ordinary mucus plugging. The casts consist of varying proportions of fibrin, mucin, and cellular material².

The clinical presentation often differs from case-to-case, with productive cough, dyspnea, and wheezing the most common findings on physical examination. Radiologic evaluation often reveals the site of the bronchial cast impaction, demonstrating atelectasis or infiltrates. Hyperinflation is often evident on the contralateral side. CT scan allows for visualization of impacted casts within the major airways. The diagnosis is usually confirmed by bronchoscopy, demonstrating airway obstruction from bronchial casts².

There are many approaches to the clinical and pathologic classification of plastic bronchitis. A pathologic approach was adopted by Seear et al.³ with a classification based on cast histology. Casts were differentiated into type I or inflammatory casts and type II or acellular casts. The type I inflammatory casts were often associated with bronchial disease. The clinical presentation of this type of cast production was often acute, rather than chronic. The acellular type II casts were found in cyanotic CHD and often in cases of idiopathic plastic bronchitis. Histologic analysis demonstrated a paucity of inflammatory cells. The acellular type of cast production was often chronic or recurrent, rather than acute. Brogan et al.⁴ classified plastic bronchitis according to associated disease states, with the following 3 categories: allergic/asthmatic, cardiac, and idiopathic. Madsen et al.¹ attempted to classify plastic bronchitis according to the underlying disease and cast morphology. The first level of classification was structural CHD¹. The patients with structural CHD were classified as mucinous, inflammatory, and chylous casts. The patients without structural heart disease, were then classified by as lymphatic disease or chylous casts, atopy or eosinophilic casts, and acute chest syndrome associated with sickle cell disease or fibrinous casts¹.

No specific therapy for bronchial casts has been shown to be effective. Different therapies have been applied, depending on the nature of the cast. Attempts to improve cardiac output in children with underlying cardiac defects are considered important. In contrast, corticosteroids have been used preferentially with inflammatory casts or type I

cast. Bronchoscopic removal has been routinely practiced in most cases^{1, 2, 4}. Presentations of plastic bronchitis with asphyxia and acute severe respiratory distress are reported and urgent bronchoscopy may be lifesaving⁶. Fragmentation or migration of the obstructing cast may induce a life-threatening event. Chest physiotherapy is also effective in promoting clearance of airway secretions. Acetylcysteine, DNase, inhaled heparin, urokinase, tissue plasminogen activator, dornase alfa, and oral macrolide antibiotics have all been used^{1, 2, 4}.

The natural history of plastic bronchitis depends on the associated disease and cast type. Patients with structural CHD are at the greatest risk of death, regardless of the cast type. The mortality in CHD associated casts is 14–50%¹. Patients who have chylous casts without CHD tend to be older and no deaths have been reported¹. There are no reported deaths in patients who have fibrinous casts, which include acute chest syndrome associated with sickle cell disease. Asthma and atopic diseases are the next most common reported association with plastic bronchitis after CHD¹. Casts associated with atopy are usually described as inflammatory with abundant eosinophils in a fibrinous background. Of the 22 reported patients with eosinophilic casts, only 12 had a defined atopic or asthmatic condition¹. The mean age was 6 years with a range from 13 months to 40 years, and there was a slight female predominance¹. This was the only group without structural CHD in which fatalities have been reported. Three of the 22 reports of patients with eosinophilic casts died due to central airway obstruction. Christensen et al.⁷ reported a 20-month-old male with presenting symptoms of cough and respiratory distress. A chest radiography showed opacification of the left lung. During bronchoscopy, he suffered cardiopulmonary arrest and anoxic encephalopathy leading to death. Microscopically, the mucus plug consisted of concentric lamellae of mucus and degenerated eosinophils. Seear et al.³ reported two deaths involving children with plastic bronchitis and eosinophilic casts. The first case was a 17-month-old boy with no history of asthma or allergy, who presented with a severe wheeze and cough following a 3 day history of upper respiratory tract infection. Bronchoscopy revealed widespread obstruction of segmental bronchi with plugs. Despite aggressive ventilation, antibiotics, corticosteroid therapy, and repeat bronchoscopy, he died 1 day after admission. Another case was a 14-year-old girl who presented with

severe mucositis 5 days after bone marrow transplantation for relapsed leukemia. Her previous respiratory health had been normal. Bronchial casts were frequently suctioned from the endotracheal tube. Despite repeated bronchoscopies, the patient died on hospital day 11. The autopsy revealed widespread airway obstruction by casts with hypocellular fibrin. This was the only group without structural CHD in which fatalities have been reported¹⁾. As we know, this is the 4th recorded case of plastic bronchitis with eosinophilic casts leading to death.

In conclusion, we have reported a child without a history of atopy, allergy, or CHD who was diagnosed with plastic bronchitis with eosinophilic casts. Plastic bronchitis can be fatal when casts obstruct the major airways, as in this case. If patients manifest signs and symptoms consistent with plastic bronchitis, clinicians should intervene early including urgent bronchoscopy and systemic treatment such as corticosteroid.

한 글 요약

건강하였던 소아에서 발생한 치명적인 호산구성 증식성 기관지염

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증식성 기관지염(plastic bronchitis)은 드문 질환으로 일반적인 점액전에 비해 크고 접착력이 있는 가지모양의 점액성 기관지 주형(cast)을 반복적으로 형성한다. 이 주형은 크기가 다양하여 자연적으로 배출되기도 하나 제거를 위해 기관지경이 필요하기도

한다. 따라서 증식성 기관지염에서 큰 기도의 폐쇄가 발생하면 생명을 위협할 수 있다. 호산구성 주형을 가진 보고된 환자 22명 중 3명이 중심 기도 폐쇄로 인해 사망에 이르렀다. 본 저자들은 아토피나 알레르기, 선천성 심장병의 병력이 없는 건강했던 소아에서 호산구성 증식성 기관지염을 진단하여 보고한다. 항생제와 스테로이드의 정주와 물리 치료를 포함한 적극적인 폐 청소와 주기적인 주형의 기관지경 제거를 시행하였으나 저산소성 뇌 손상에 이어 뇌사에 이르렀다. 본 증례와 같이 증식성 기관지염이 중심 기도를 막았을 때 생명을 위협할 수 있다. 따라서 증식성 기관지염의 증상이나 징후가 보일 경우 조기 치료가 필요하리라 생각 된다.

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