Korean Journal of Pediatrics Vol. 52, No. 7, 2009
DOI : 10.3345/kjp.2009.52.7.832
■ Case report ■

Plastic bronchitis in children: 2 cases

Yeo Hyang Kim, M.D., Hee Jung Choi, M.D.*
Jung Ok Kim, M.D.† and Myung Chul Hyun, M.D.‡

Department of Pediatrics, School of Medicine, Keimyung University, Daegu, Korea
Department of Pediatrics*, Kumi-Cha Hospital, Kumi, Korea
Division of Pediatric Cardiology†, Department of Pediatric, Severance Cadiovascular Hospital
Yonsei University College of Medicine, Seoul, Korea
Departments of Pediatrics‡, School of Medicine, Kyungpook National University, Daegu, Korea

= Abstract =

Plastic bronchitis is a rare disorder characterized by the formation of extensive, obstructing endobronchial casts. It is associated with asthma and complex cardiac defects such as those requiring the Fontan procedure. The treatment of plastic bronchitis comprises conventional therapy involving spontaneous expectoration and bronchoscopic removal and specific therapy with several new drugs. Herein, we describe the cases of 2 patients diagnosed with plastic bronchitis accompanied with a different underlying disease, which were treated with inhaled corticosteroid and low-dose oral clarithromycin. **(Korean J Pediatr 2009; 52:832-836)**

Key Words : Plastic bronchitis, Children, Congenital heart disease, Bronchoscope

Introduction

Plastic bronchitis is an uncommon disorder characterized by the recurrent formation of mucoid bronchial casts. It is usually associated with primary pulmonary diseases including asthma, cystic fibrosis, allergic bronchopulmonary aspergillosis, and congenital heart defects[1]. In Korea, a few cases with different underlying disease have been reported since 1995[2-4]. Common complaints are cough, fever, and dyspnea. The most common finding on physical examination is wheezing, and radiologic findings show hyperinflation due to air trapping or atelectasis secondary to airway obstruction by bronchial cast[5]. The treatment of plastic bronchitis includes therapy for cast removal or expectoration, and management for the underlying pulmonary and cardiac disease[5]. In some cases, anti-inflammatory medications such as inhaled corticosteroids or macrolide antibiotics are useful[6, 7]. We describe two patients diagnosed as plastic bronchitis with different underlying disease, which were treated with inhaled corticosteroid and low dose oral clarithromycin.

Received : 22 October 2008, Revised : 15 April 2009
Accepted : 11 June 2009
Address for correspondence : Myung Chul Hyun, M.D., Ph.D.
Department of Pediatrics, Kyungpook National University Hospital, 200 Dongduk-Ro, Jung-Gu, Daegu, 700-721 Korea
Tel : +82.53-420-5704, Fax : +82.53-425-6683
E-mail : mchyun@mail.knu.ac.kr
*These cases were presented in the Annual Meeting of the European Respiratory Society in Berlin, Germany, October 4-8, 2008.

Case report

1. Case 1

An 8-year-old boy was referred for foreign body removal using rigid bronchoscopy to our center. He had been admitted to other hospital 2 days before for productive cough, fever, and chest radiographic findings of total haziness of left lung (Fig. 1A). Diagnosis on admission was pneumonia and he was treated with anti-tussives, mucolytics, and antibiotics. On the following day, his condition suddenly deteriorated with tachypnea, hypoxia, retracting respiration with diffuse wheezing and poor aeration of left lung. At that time, chest CT revealed low density materials in left bronchus. Foreign body aspiration in left bronchus was suspected and he was referred for bronchoscopy to our center. His personal past history and family history were unremarkable.

On admission day to our center, his body temperature was 38.7℃, heart rate 130/min, respiratory rate 30/min and blood pressure 115/59 mmHg. On physical examination, he had acute ill appearance with tachypnea, wheezing and poor

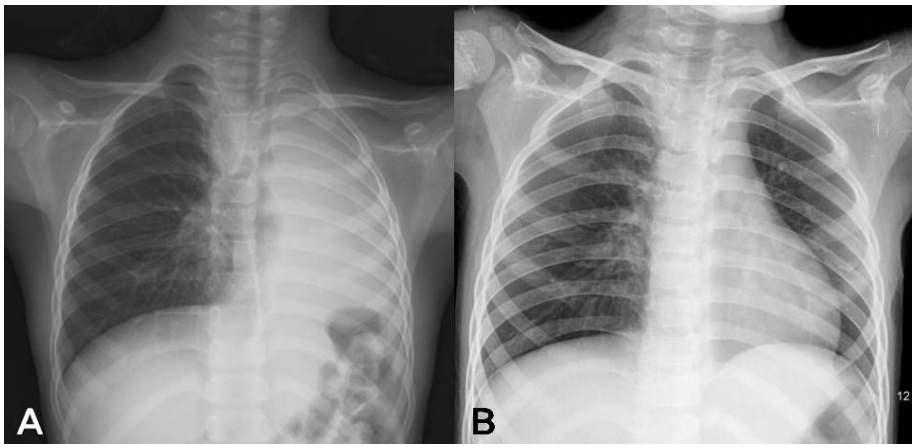

Fig. 1. Improvement after bronchoscopic removal in case 1. (A) Initial chest radiograph showing complete atelectasis of the left lung. (B) Follow-up chest radiograph showing complete resolution after bronchoscopic removal and specific therapy with inhaled corticosteroid and low-dose macrolide.

aeration in left lung field. A complete blood count showed white blood cell count 4,990/mL (neutrophil 88%; lymphocyte 7%; monocyte 1.9%), hematocrit 36% and platelets 420,000/mL. Arterial blood gas analysis (ABGA) on oxygen mask 6L inhalation showed pH 7.48 PCO_2 30 mmHg PO_2 84 mmHg saturation 97%. The patient was presented to the operating room for emergency diagnostic bronchoscopy. He was in the supine position and ventilating rigid bronchoscopy with general anesthesia was performed. On bronchoscopic examination, mucoid material from left main bronchus was found and removed to pieces using forceps. Although complete removal could not be done because of bronchial swelling, breathing sound and aeration of left lung field were improved after the procedure.

On the third admission day, he had more severe retracting respirations, tachypnea and generalized wheezing with poor aeration. A repeat chest X-ray showed hyperinflation but little change from previous studies. ABGA showed hypoxia and retention of PCO_2 as 80 mmHg. Endotracheal intubation was done and thick yellow sputum came from trachea during endotracheal suction. Treatments included intravenous systemic corticosteroid, theophyllin and broad spectrum antibiotics. With these treatments, his clinical symptoms and condition seemed to be improved. A follow-up laboratory test showed white blood count 16,600/mL (neutrophil 81%; lymphocyte 9%; monocyte 3.4%), hematocrit 35% and platelets 523,000/mL. Urine pneumococcal antigen, cold agglutinin and mycoplasma antibody (IgM and IgG) were all negative findings. Specific IgE test using allergic MAST and unicap was negative. Eosinophil cationic protein was 18 ng/mL, eosinophil was 5.6% and total IgE was 5,000 IU/L.

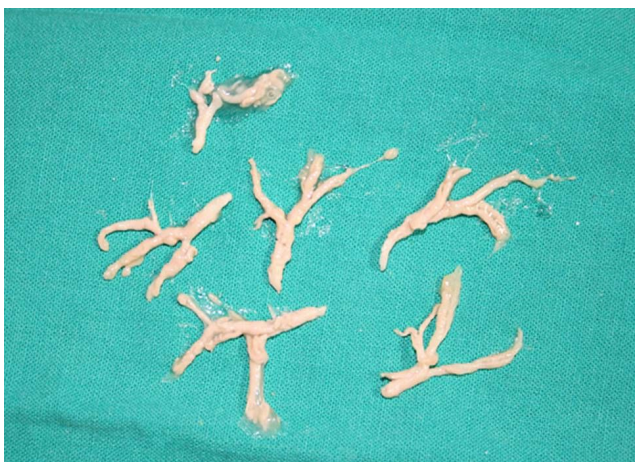

Fig. 2. Yellow thick and gelatinous fragments of bronchial cast, which were removed from the left bronchus by flexible bronchoscophy in case 1.

On the fourth admission day, a repeat chest X-ray showed left lung total haziness and aeration was decreased at left lung field. Thick yellow endotracheal excretions were persistent.

In spite of treatment with inhaled beta agonist, anticholinergics, steroid, antibiotics and massive physiotherapy, respiratory distress was worsening and opacification of left lung was repeated. Evaluation using further bronchoscopy for airway obstruction was decided. On the flexible bronchoscopic examination, complete occlusion of the left upper and lower bronchi by yellow thick materials was noted and large gelatinous branching airway cast was removed (Fig. 2). Microscopic evaluation found the cast consisted of muco-

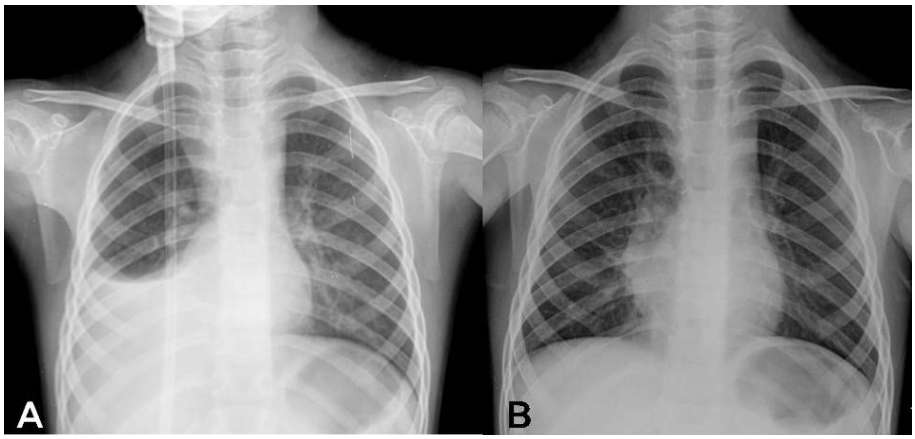

Fig. 3. Improvement after spontaneous expectoration in case 2. (A) Initial chest radiograph showing bilateral perihilar consolidation and right middle and lower lung collapse. (B) Follow-up chest radiograph showing complete resolution after spontaneous expectoration.

fibrin with eosinophils, lymphocytes and plasma cells. All cultures were negative. And finally, plastic bronchitis was diagnosed.

During the 6 months following procedure, the patient has used inhaled steroid with low dose oral macrolide and had no further episodes of respiratory distress or cast formation (Fig. 1B).

2. Case 2

A 3-year-old girl presented to our center with a 2-day-history of productive coughing, increased work of breathing, facial cyanosis and cold sweating.

At her past medical history, she had undergone multiple cardio-thoracic procedures including fenestrated Fontan operation with an intracardiac tunnel for criss cross heart, transposition of the great artery, pulmonary atresia and ventricular septal defect. 2 month later postoperatively, she developed bilateral pleural effusion and was diagnosed as chylothorax. She needed operation for thoracic duct ligation and was discharged after operation with warfarin and angiotensin converting enzyme inhibitors.

Her acute respiratory distress with hypoxia developed 1 year after the Fontan operation. Her physical exam on admission was afebrile state with a respiratory rate 70/min, oxygen saturation 73% and PaO_2 38 mmHg on room air. Auscultation of her chest revealed decreased breath sound in right lung field. Chest radiograph demonstrated an increase in bilateral interstitial infiltration and right-side atelectasis (Fig. 3A). Little clinical improvement was made in spite of medical therapy. On the third admission day when she had

Fig. 4. A whitish, tree-like bronchial cast that was spontaneously expectorated in case 2.

post-tussive emesis, she expectorated an elongated whitish soft tissue of the bronchial tree (Fig. 4) and her symptom was resolved (Fig. 3B). Microscopic evaluation of the expectorated tissue found the cast consisted of mucofibrin with a few acute inflammatory cells. Eosinophil or Charcot-Leyden crystal was not found and all cultures were negative. Plastic bronchitis was diagnosed.

Respiratory distress with hypoxia recurred 2 weeks later. She was treated with massive physiotherapy, inhaled corticosteroid, bronchodilator, mucolytics and broad spectrum antibiotics. Like previous admission, she repeatedly expectorated a bronchial cast and her symptom was resolved.

Respiratory distress recurred within 2 weeks secondary to bronchial cast in the right lung. Rigid bronchoscopy was performed and complete occlusion of the right main bronchus

by thick materials was noted and large gelatinous branching airway cast was removed.

Echocardiogram showed good patent Fontan circulation without enlargement of venous system but cardiac catheterization could not be performed due to failure of wire introduction into both femoral veins and right jugular vein.

During the 6 months following procedure, the patient had sixth episodes of respiratory distress with cast formation and has used regularly inhaled steroid with low dose oral macrolide. In 9 months since regular medication, the patient has not had further respiratory difficulty or cast reaccumulation.

Discussion

According to pathological classification by Seear[6], bronchial casts were differentiated into type I (cellular or inflammatory) or type II (acellular). Type I casts occur in patients with underlying pulmonary inflammatory disease and type II casts develop more commonly in patients undergone surgery for cyanotic heart disease.

Our first patient had no definite underlying pulmonary pathology. Although IgE level was very high, methacholine challenge test was not performed for this patient and there was no clear evidence that he had had asthma in the past and personal history. Our second patient underwent Fontan procedure and thoracic duct ligation. Although acellular casts are more common type for congenital heart disease, our two patients showed pathological findings consisted of inflammatory cast.

The first treatment of plastic bronchitis is removal of casts by bronchoscope or spontaneous expectoration. In addition, chest physiotherapy and several mucolytic agents have been used[5, 8]. Although appropriate removals of large casts resolve airway obstruction, long-term therapy for the underlying disease should be initiated for prevention of cast recurrence. In particular, use of anti-inflammatory medications can be helpful for type I inflammatory cast bronchitis.

Corticosteroids as anti-inflammatory agents have been used in plastic bronchitis[6, 9]. Wang et al reported in a retrospective clinical trial study of adult plastic bronchitis with hemoptysis[10]. They demonstrated that routine doses of corticosteroids as an anti-inflammatory drug would be effective and safe treatment of plastic bronchitis with hemoptysis and recommended early intervention with corticosteroids when bronchial casts were showed.

The 14- and 15-member macrolide antibiotics are drugs with both immunomodulatory and mucoregulatory effects, unrelated to antimicrobial properties[11]. There are several evidences suggesting the usefulness of macrolide antibiotics, as immunomodulating agent, in the treatment of other inflammatory pulmonary disease including diffuse panbronchiolitis (DPB)[12], cystic fibrosis[13], asthma, and plastic bronchitis[7]. In Japan, erythromycin has been used in the treatment of DPB[12]. DPB shows recurrent cough, productive sputum, wheeze, and exertional dyspnea. In this disease, chronic airway obstruction and infection cause to progressive bronchiectasis, impairment of pulmonary function, and respiratory failure. Recently, since the macrolides are used for treatment of DPB, 10-year-survival of DPB has been increased from 12.4% to over 90%. Schultz et al reported first experience of the use of azithromycin for plastic bronchitis[7]. This treatment resulted in normalization of the patients spirometric measurements and improvement of his clinical symptoms. They concluded that azithromycin was more effective than either bronchoscopy or inhaled steroids. Previous reports demonstrated that the anti-inflammatory properties by macrolide antibiotics rather than their antimicrobial properties were related to beneficial effects.

Our patients showed pathological findings consisted of inflammatory cast, so we used long-term therapy with inhaled corticosteroids and low dose oral clarithromycin. All of them did not show symptoms or signs of bronchial obstruction by recurrent cast formation after using those medications. We do not know the exact pathophysiology of cast formation in our patients, but suspect the association with inflammation. The pathologic finding and clinical response to anti-inflammatory treatments may be evidence for that hypothesis of disease pathology. Therefore, we recommend early intervention with anti-inflammatory drugs in addition to conventional therapy if the patients with plastic bronchitis show sufficient clinical evidence.

한 글 요 약

소아 증식성 기관지염 2례

*계명대학교 의과대학 소아과학교실, 구미차병원 소아과청소년과**
연세대학교 의과대학 심장혈관병원 소아심장과†
경북대학교 의과대학 소아과학교실‡

김여향 · 최희정* · 김정옥† · 현명철‡

증식성 기관지염(plastic bronchitis)는 기관지 내부에 폐쇄성 원주(cast)를 만드는 드문 질환으로 천식이나 선천성 심장병의

폰탄 수술 후 합병증으로 생길 수 있다. 증식성 기관지염의 치료는 원주를 스스로 뱉거나 기관지경으로 제거해주는 고식적인 치료와 새로 개발된 약제를 사용하는 단기 및 장기 치료로 나눌 수 있다. 저자들은 기저 질환이 없이 갑작스런 호흡 부전과 왼쪽 폐 전체의 무기폐를 보인 8세 남아와 폰탄 수술 병력이 있고 반복되는 오른쪽 중엽과 하엽의 무기폐를 보인 5세 여아에서 기관지 내시경을 통해 기관지내 원주를 확인하였고, 조직학적 검사에서 호산구, 호중구, 임파구와 같은 염증 세포를 포함한 점액질과 섬유질로 구성되어 있어 증식성 기관지염으로 진단후 재발 방지를 위해 흡입용 스테로이드와 저용량 경구용 clarithromycin을 사용한 경험을 문헌고찰과 함께 보고하는 바이다.

References

1) Madsen P, Shah SA, Rubin BK. Plastic bronchitis: new insights and a classification scheme. Paediatr Respir Rev 2005;6:292-300.
2) Kim CH, Kim SJ, Kim YK, Lee YJ, Kim KE, Lee KY. A. case of bronchial cast. Pediatr Allergy Respir Dis (Korea) 1996;6:172-6.
3) Kim YS, You JH, Rhee KS, Rha KS. A Case of Bronchial Cast. Korean J Pediatr 1988;31:1232-6.
4) Goo HW, Jhang WK, Kim YH, Ko JK, Park IS, Park JJ, et al. CT findings of plastic bronchitis in children after a Fontan operation. Pediatr Radiol 2008;38:989-93.
5) Eberlein MH, Drummond MB, Haponik EF. Plastic bronchitis: a management challenge. Am J Med Sci 2008;335:163-9.
6) Seear M, Hui H, Magee F, Bohn D, Cutz E. Bronchial casts in children: a proposed classification based on nine cases and a review of the literature. Am J Respir Crit Care Med 1997;155:364-70.
7) Schultz KD, Oermann CM. Treatment of cast bronchitis with low-dose oral azithromycin. Pediatr Pulmonol 2003;35:135-43.
8) Nayar S, Parmar R, Kulkarni S, Cherian KM. Treatment of Plastic Bronchitis. Ann Thorac Surg 2007;83:1884-6.
9) Onoue Y, Adachi Y, Ichida F, Miyawaki T. Effective use of corticosteroid in a child with life-threatening plastic bronchitis after Fontan operation. Pediatr Int 2003;45:107-9.
10) Wang G, Wang YJ, Luo FM, Wang L, Jiang LL, Wang L, et al. Effective use of corticosteroids in treatment of plastic bronchitis with hemoptysis in Chinese adults. Acta Pharmacologica Sinica 2006;27:1206-12.
11) Shinkai M, Rubin BK. Macrolides and airway inflammation in children. Paediatr Respir Rev 2005;6:227-35.
12) Keicho N, Kudoh S. Diffuse panbronchiolitis: role of macrolides in therapy. Am J Respir Med 2002;1:119-31.
13) Wolter J, Seeney S, Bell S, Bowler S, Masel P, McCormack J. Effect of long term treatment with azithromycin on disease parameters in cystic fibrosis: a randomized trial. Thorax 2002;57:212-6.