Case Report

Congenital Lobar Emphysema in a Toddler: The Role of Flexible Bronchoscopy in Diagnosis and Treatment

Shu-Leei Tey[1], Chien-Yi Wu[1], Yu-Tsun Su[1], Chia-Chang Tsai[1], Yung-Cheng Lin[1]

Congenital lobar emphysema (CLE) with respiratory distress is traditionally thought to be best treated by lobectomy. However, mild-to-moderate cases of CLE are generally treated conservatively in recent years with the increasing use of either flexible or rigid bronchoscopy to locate and treat some of the associated airway anomalies. On the other hand, there is no consensus regarding the role of flexible bronchoscopy (FB) in the diagnosis and treatment of CLE. Herein we report a case of CLE in a 2-year 4-month-old boy with moderate respiratory distress, who was treated conservatively with success. A tracheal bronchus with bronchomalacia at the orifice of the right upper lobe was diagnosed by FB. A review of literature regarding the use of bronchoscopy as a treatment strategy for CLE is also presented. FB can be used in patient with CLE to better identify the etiologies and determine whether to treat the patients conservatively or to perform lobectomy.

Key words: bronchomalacia, bronchoscopy, pediatrics, pulmonary emphysema, tracheal bronchus

Introduction

Congenital lobar emphysema (CLE) is characterized by over-inflation and air-entrapment in the affected lobe or lobes.[1] This congenital malformation is rare, with a prevalence of 1 in 20,000 to 30,000.[2] It usually appears during infancy and early childhood, and the symptoms vary from asymptomatic to severe cyanosis. Its causes involve intrinsic or extrinsic factors leading to bronchial obstruction and alveolar over-distention.[3] Bronchial cartilage dysplasia has been reported to be the cause of CLE in 50% to 71% of the cases, while other contributing factors also include vascular compression, mucosal thickening, and granulation tissues.[1,4] Although lobectomy was once considered to be the best treatment for CLE with respiratory distress, conservative

From the [1]Department of Pediatrics E-Da Hospital, I-Shou University, Kaohsiung, Taiwan.
Received: April 23, 2012 Accepted: October 1, 2012
Address reprint request and correspondence to: Yu-Tsun Su, Department of Pediatric Pulmonology, Department of Pediatrics, E-Da Hospital, No. 1, Yida Road, Jiaosu Village, Yanchao District, Kaohsiung City 82445, Taiwan.
Tel: +886-7-6150011 ext. 251072, Fax: +886-7-6150950, E-mail: ed100616@edah.org.tw

treatment is increasingly suggested for patients with mild-to-moderate symptoms.[1,3,5-7] Chest X-ray (CXR), computed tomography (CT), multislice computed tomography (MSCT)[8-9] with virtual bronchoscopy,[8] and perfusion scan are often used to diagnose CLE, whereas flexible bronchoscopy (FB) is being used more frequently to observe its associated airway anomalies.[3,5-6,10-12] MSCT is used to evaluate the location and associated anomalies of CLE in recent.[8-9] Nevertheless, there is still no consensus about the role of FB in the diagnosis and treatment of CLE. We discuss the role of FB in the diagnosis of CLE and selection of treatment strategy by reporting a 2-year 4-month-old boy being successfully treated conservatively and reviewing the relevant literature.

Case Report

A 2-year 4-month-old boy had been born prematurely (GA 33 weeks, BBW 1,920 gm). He was intubated for two days after birth under the impression of respiratory distress syndrome Grade II without surfactant treatment. The CXR on Day 1 and Day 3 revealed bilateral mild infiltration without lobar emphysematous change. He had an uneventful course of recovery and was discharged when he was thirty days old. When he was 1.5 years old, he was found by CXR to have right lobar emphysematous change. From that time, he had had several episodes of wheezing and respiratory tract infection. When 2-year 4-month-old, he was admitted to a regional hospital for tachypnea and bilateral wheezing that had lasted for more than one week. The CXR (Fig. 1), high resolution computed tomography (HRCT) with reconstruction of tracheobronchial tree, and virtual bronchoscopy revealed emphysematous change of right upper lobe and a suspicious focal nodular protrusion causing narrowing of right upper bronchial lumen without any cardiovascular anomaly. Though he was not reported to have chocking, foreign body aspiration was suspected.

He was referred to our hospital and admitted to our pediatric intensive care unit due to tachypnea with suprasternal retraction and apparent bilateral wheezing. His respiratory rate was 40 per minute and oxygen saturation 95% under fraction of inspired oxygen (FiO2) 35%. Perfusion scan revealed mildly decreased perfusion over the right upper lobe. FB (Olympus BF-N20, 2.2 mm diameter) showed bronchomalacia at the orifice of the tracheal bronchus to the right upper lobe, with patency during inspiration (Fig. 2) and dynamic collapse during expiration (Fig. 3). Advancing the flexible bronchoscope through the malacic portion revealed 3 normal segmental bronchi. Based on FB, the patient was diagnosed as having CLE resulting from bronchomalacia of tracheal bronchus due to bronchial cartilage defect. His wheezing improved after bronchodilator therapy, although the breath sounds over right upper lung field were slightly decreased. He was discharged after five days. Conservative treatment for CLE was suggested and Fluticasone Evohalor was prescribed for mild persistent asthma. The child has been well for 18 months of follow-ups.

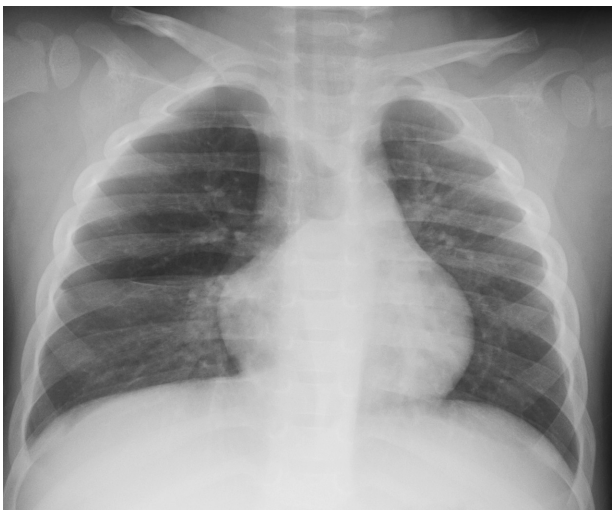

Fig. 1 Chest radiography showing emphysematous change of right upper lobe with contralateral herniation.

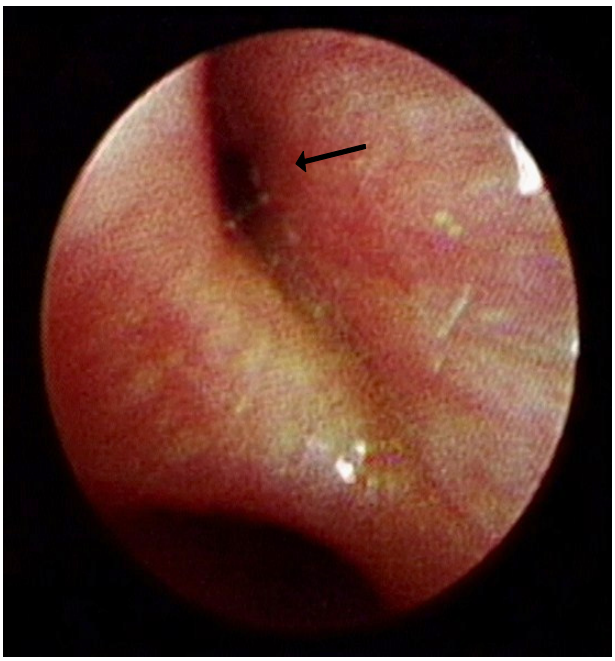

Fig. 2 Bronchomalacia of the tracheal bronchus: patency during the inspiration phase. (Black arrow: the orifice of the tracheal bronchus to the right upper lobe.)

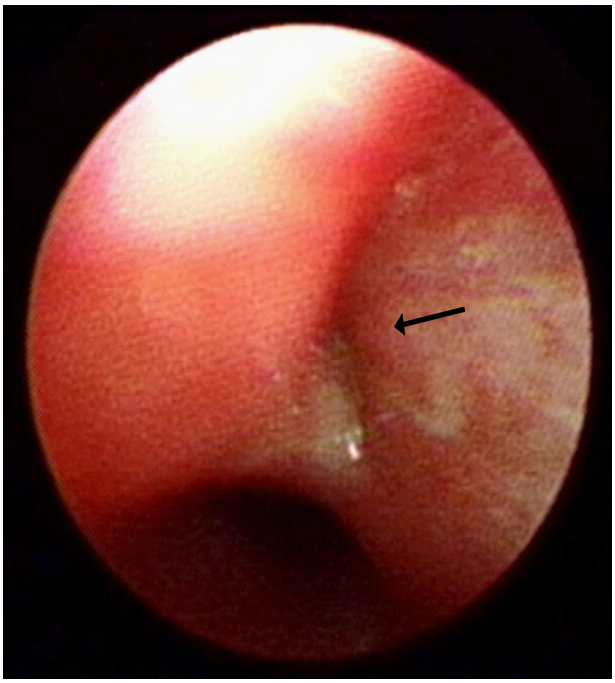

Fig. 3 Bronchomalacia of the tracheal bronchus: complete collapse during the expiration phase. (Black arrow: the orifice of the tracheal bronchus to the right upper lobe.)

Discussion

In CLE with respiratory distress, lobectomy was once considered to be the best treatment regardless of the diverse etiologies. Recently, with the use of FB, more cases of CLE have been found to be associated with airway anomalies, and conservative treatment has been increasingly used for cases with mild-to-moderate symptoms.[1,3,5-7,10-12] Many patients with CLE who underwent FB for the diagnose of bronchomalacia have been treated conservatively. Other bronchoscopic interventions, such as polypectomy and stenting, had achieved favorable outcomes comparable to those from emergent lobectomy for CLE.[11-13] In this case, we used FB to diagnose bronchomalacia in a 2-year 4-month-old toddler with CLE, who was treated conservatively and has been well for 18 months of follow-ups.

The role of FB in diagnosis and conservative treatment of CLE associated with bronchomalacia

FB can evaluate dynamic changes in the airway, including bronchomalacia and external vascular compression.[14] We searched the Pubmed database using the keywords “bronchoscopy” and “congenital lobar emphysema” for English literature published from 1991 to 2012 (Table 1). Bronchomalacia, which is a bronchoscopic finding in many patients with CLE presenting with different respiratory problems, has been treated either conservatively or by lobectomy. FB began to gain popularity among pediatricians after the first publication endorsing the use of flexible bronchoscope as an effective tool for exploration of the pediatric airway in 1984.[15] After that, expert teams have reported no absolute contraindication and also a low complication rate for the procedure. The relative contraindications are a tendency toward bleeding, severe airway narrowing, and severe impairment of cardiopulmonary functions.[15] FB had been increasingly performed as a standard procedure to identify reversible causes, such as foreign body aspiration, in patients with suspected CLE. Ozcelik et al. studied twenty-

Table 1. Reviewed cases reporting the use of bronchoscopy with congenital lobar emphysema from 1991 to 2012 (n/N = abnormal cases/all cases)

Author	CLE cases	X-ray	CT	Perfusion scan	Bronchoscopy (flexible or rigid)	Bronchoscopic finding	Treatment	Pathology
Hochheggar et al. 2012[8]	1	1	1	0	0	0	Lobectomy	Bronchial stenosis
Nath et al. 2011[2]	1	1	1	0	0	0	Lobectomy	N/A
Kovacevic et al. 2009[5]	2	2/2	2/2	1/1	2/2 (flexible)	Diffuse bronchomalacia (1/2) Bronchial stenosis (1/2)	Conservative	N/A
Clubley et al. 2007[11]	1	1/1	0	0	1/1 (rigid)	Pedunculated polyp over bronchus	Removed by optical grasping forceps	Granulation tissue
Seo et al. 2006[19]	2	2/2	2/2	0	2/2 (CT bronchoscopy)	Bronchial atresia (2/2)	Lobectomy (2/2)	Maldevelopment of the bronchial cartilage (1/2) Hypertrophied, fused bronchial cartilages (1/2)
Chao et al. 2005[10]	1	1/1	1/1	0	1/1 (bronchoscopy in OR)	Bronchomalaica (1/1)	Lobectomy	N/A
Shanmugam et al. 2005[20]	9	9/9	N/A	0	4/9	N/A	Lobectomy	N/A
Ozcelik et al. 2003[3]	30	30/30	16/16	8/8	0/7 (rigid) 1/1 (flexible)	Narrowed and flaccid bronchus (1/ 8)	Lobectomy (21/30) Conservative (9/30)	Emphysema (21/21) Bronchial cartilage deficiency (2/21)
Karnak et al. 1999[6]	14	14/14	12/12	7/8	2/6	Serous secretion (1) Bronchial stenosis (1) Normal (4/6)	Lobectomy (10/14) Conservative (4/14)	Hypoplastic cartilage (2/10) No specific finding (8/10)
Phillipos and Libsekal 1998[12]	1	1/1	0	0	1/1 (flexible)	Bronchomalacia (1/1)	Lobectomy (1/1)	Bronchial cartilage hypoplasia (1/1)
# Mikhailova 1996[18]	32	usually	8/(N/A)	N/A	*8/(N/A)	*Bronchial stenosis (8/(N/A))	Lobectomy (32/32)	Cartilage deficit (14/32), Bronchial stenosis (26.2%), Unclear (43.8%)
Doull et al. 1996[16]	1	1/1	0	@1/1	1/1 (flexible)	Bronchomalacia	Conservative (1/1)	N/A
Saim et al. 1994[4]	1	1/1	0	0	Bronchoscopy	Bronchial septum (1/1)	Lobectomy initial, perforate the membranous septum and dilate the perforation	Distended alveoli, distention and rupture of the terminal bronchus
Stigers et al. 1992[21]	8	8/8	4/4	@2/2	Bronchoscopy (2/3) (all conservative treatment)	Bronchial stenosis (2/3)	Lobectomy (3/8), Medical treatment (5/8)	Hypoplasia of peripheral bonch (1/5) Alveolar rupture and distention (2/3)

CLE: Congenital lobar emphysema; CT: Computed tomography; @: Ventilation/perfusion scan;
* Stenosis of the respective lobar bronchus is discovered in 8 children by CT and bronchoscopy.
# Article in Bulgarian; N/A: Not available

one patients with CLE of whom twelve were treated by lobectomy and nine were treated conservatively. CXR follow-up revealed improvement in hyperlucency of the affected lobe in the conservatively treated group.[3] The most common cause of CLE is bronchomalacia resulting from bronchial cartilaginous dysplasia or deficiency, which presents as airway patency during inspiration and collapse during expiration.[3] Conservative treatment may be appropriate in patients with CLE associated with bronchial stenosis or bronchomalacia presenting with mild-to-moderate symptoms. Kovacevic et al. reported two patients with CLE receiving FB and conservative treatment. The first case was one of bronchomalacia and hyperlucency of the left upper lobe which had almost disappeared on CXR follow-up, most likely due to maturation and growth of bronchial cartilage. The second patient, who had bronchial stenosis, was almost symptom-free after treatment.[5] Doull et al. reported a 3-year-old boy with CLE who was found to have bronchomalacia of left apico-posterior segmental bronchus by FB which showed evidence of pulmonary over-inflation attributable to check valve effect.[16] The authors suggested that FB was useful in the evaluation of airway patency and dynamic change in CLE.[5,16] Conservative treatment and close follow-up are appropriate for patients with CLE presenting with mild-to-moderate symptoms, especially those with bronchomalacia.

The role of FB in non-surgical intervention and lobectomy

Bronchoscopic findings suggest that some non-surgical interventions provide favorable outcomes comparable to those through emergent lobectomy for patients with CLE. Using bronchoscopy, Clubley et al. reported an infant with lobar emphysema caused by a pedunculated polyp prolapsing in and out of the right bronchus intermedius. Optical grasping forceps were used to remove the polyp from the infant, whose post-operative course was smooth with no need for lobectomy.[11] In a report by Ide et al., a one-month-old boy with severe emphysema of right upper lobe due to stenotic tracheal bronchus compressed by vascular anomalies received a cardiovascular operation that failed to alleviate his severe hypoxia due to persistent bronchomalacia of the tracheal bronchus for which bronchoscopic stenting was performed and the patient was eventually weaned from mechanical ventialtion.[13] Phillipos et al. reported a neonate with bronchomalacia and CLE of left upper lobe. The baby's initial presentation on CXR and respiratory distress improved after introduction of an ultrathin FB into the LUL bronchus to relieve the check valve effect. The neonate received elective lobectomy 3 days later. In that report, the authors proposed that FB may relieve lobar hyperinflation of the lung secondary to bronchial cartilage hypoplasia or segmental bronchomalacia.[12] These reports underscore the importance of bronchoscopy prior to lobectomy in patients with suspected CLE. Based on these reports, FB may be used as a diagnostic tool as well as a means of selective treatment for patient with CLE to avoid unnecessary lobectomies[5-6] that can be reserved for those with severe respiratory distress.[3,6,17-18]

Conclusions

Though the etiologies of congenital lobar emphysema (CLE) are diverse, more than fifty percent of the cases have been reported to be caused by cartilage dysplasia, presenting solely as bronchial stenosis and bronchomalacia. Since FB can directly and dynamically evaluate the airway anomalies in CLE, it can be utilized as a standard procedure to identify reversible causes in patients with suspected CLE. Bronchomalacia-related CLE with mild-to-moderate symptoms can be treated conservatively with close follow-ups, whereas lobectomies should be reserved for CLE with severe

respiratory distress. FB should be used to study suspected CLE cases to determine their causes and decide whether to treat the patient conservatively, by lobectomy, or by other strategies. The use of FB, therefore, may reduce the number of unnecessary surgical procedures.

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