International Journal of Pediatric Otorhinolaryngology (2005) **69**, 549—554

ELSEVIER

International Journal of Pediatric Otorhinolaryngology

www.elsevier.com/locate/ijporl

CASE REPORT

Congenital lobar emphysema: an Otolaryngologic perspective

Michael C. Chao*, **Amir M. Karamzadeh, Gupreet Ahuja**

Division of Pediatric Otolaryngology, Department of Otolaryngology—Head and Neck Surgery, University of California Irvine Medical Center, 101 The City Drive, B-25 R-81, Orange, CA 92868, USA

Received 28 April 2004; received in revised form 15 October 2004; accepted 6 November 2004

KEYWORDS
Congenital lobar emphysema;
Respiratory distress;
Endoscopy;
Infant

Summary Congenital lobar emphysema (CLE) is an uncommon but potentially life threatening pulmonary abnormality affecting infants. Patients often present within the first 6 months of life with recurrent respiratory distress as a result of hyperinflation of the affected pulmonary lobe, and resultant near total collapse of normal lung parenchyma. We present a case of a 2-month-old infant with recurrent admissions for respiratory distress.

1. Introduction

Congenital lobar emphysema (CLE) is a rare cause of airway compromise affecting the pediatric age group. Disease presentation can vary from acute respiratory distress to recurrent bouts of pneumonia. Radiographically, CLE is characterized by hyperinflation of the affected pulmonary lobe. Diagnosing this rare entity amidst the more come common pulmonary and airway pathologies poses a significant challenge. The Otolaryngologist may be consulted to assess the status of the airway and help rule out intraluminal etiologies, such as foreign bodies, which present in a similar fashion. Given the tenuous nature of the airway in certain affected individuals, it is very important to keep this entity in mind before performing bronchoscopy. We present a case of CLE and review the presentation, diagnostic evaluation, and highlight the otolarygologic considerations.

2. Case report

We present a 2-month-old male who has a history of two admissions to the pediatric intensive care unit at a community hospital for respiratory distress and pneumonia which required intubation. On the third admission, the patient was found to be in marked respiratory distress and hypoxic, with oxygen saturation in the low 90s, while breathing room air. He was transferred to UC Irvine Medical Center. On physical examination, the patient was noted to have marked substernal retractions and usage of accessory respiratory muscles. Interestingly, the patient's air exchange and work of breathing improved when placed in the left lateral decubitus position. Chest röentgenogram on admission sug-

* Corresponding author. Tel.: +1 714 456 5753; fax: +1 714 456 5747.
E-mail address: mcchao@uci.edu (M.C. Chao).

0165-5876/$ — see front matter
doi:10.1016/j.ijporl.2004.11.017

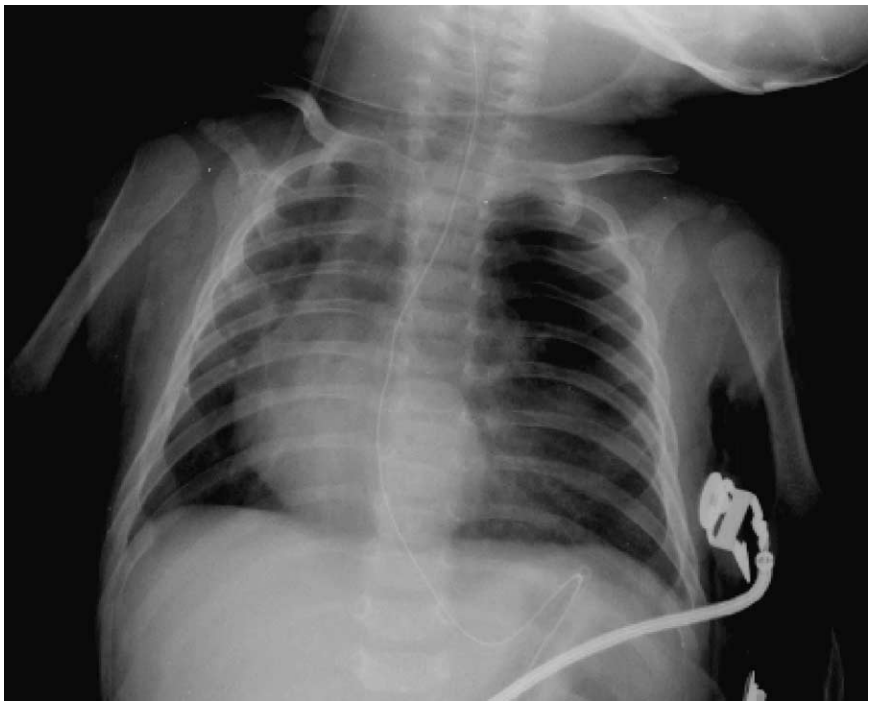

Fig. 1 AP chest röentgenogram. Note the hyperinflated left lung with the mediastinum shifted to the right.

gested hyperinflation of the left lung and mediastinal deviation to the right (Fig. 1).

The Otolaryngology service was consulted to assess the airway. The patient was, subsequently taken to the operating room to undergo diagnostic direct laryngoscopy and bronchoscopy for presumptive left lung and bronchial pathology. Intraoperatively, the patient was noted to have a normal glottis and subglottis with mobile true vocal cords. There was no evidence of laryngomalacia or tracheomalacia. The right mainstem bronchus and bronchioles were patent and free of abnormality. The left lower lobe bronchus was noted to be dynamically compressed, with critical stenosis on inspiration and complete collapse on expiration (Figs. 2 and 3); no foreign body or intraluminal pathology was noted. The patient was intubated after direct laryngoscopy and bronchoscopy were performed.

Subsequent radiographic examinations included computed tomography (CT), magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of the chest (Figs. 4 and 5). While the CT chest was negative for any intrathoracic masses it did

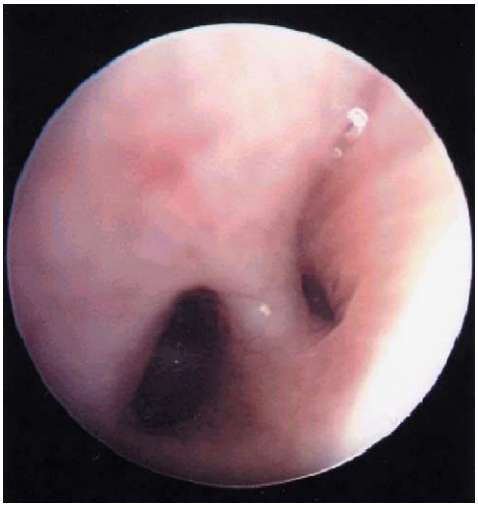

Fig. 2 Bronchoscopic evaluation of the left main bronchus demonstrating stenotic left lower bronchus on inspiration.

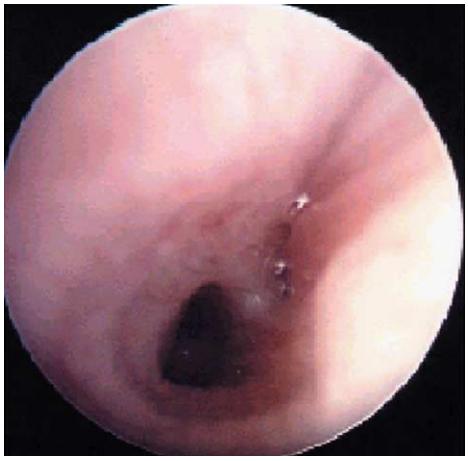

Fig. 3 Bronchoscopic evaluation of the left main bronchus demonstrating complete collapse of the left lower bronchus on expiration.