International Journal of Current Advanced Research
ISSN: O: 2319-6475, ISSN: P: 2319 – 6505, Impact Factor: SJIF: 5.438
Available Online at www.journalijcar.org
Volume 6; Issue 1; January 2017; Page No. 1759-1761

Case Report

CONGENITAL LOBAR EMPHYSEMA IN AN ADOLESCENT

Yadav Sambhav., Taneja vipul and Bhardwaj AK

Departments of Pediatrics, MMIMSR , Mullana, Ambala, Haryana, India

ARTICLE INFO

Article History:

Received 9th October, 2016
Received in revised form 27thNovember, 2016
Accepted 4th December, 2016
Published online 28th January, 2017

Key words:

Adolescent male, CLE, severe breathlessness.

ABSTRACT

Congenital lobar emphysema (CLE) is a clinico-radiological diagnosis, seen usually by four-six weeks of age (50% of patients) and rarely (<5% of patients) after the age of six months. Here, we report a adolescent male with gradual onset of severe breathlessness and physical examination revealing the features of Bronchial Asthma. X-ray of chest, with subsequent CT of chest, leads to the diagnosis of CLE. The pulmonary function tests, bronchoscopic examination and α1-antitrypsin level are normal. Patient is managed conservatively.

INTRODUCTION

Developmental lung anomalies in the adolescent can be a challenge, as the abnormality may be mistaken for something more sinister. Congenital lobar emphysema (CLE) is a lung bud (bronchopulmonary) anomaly, usually detected in the neonatal period and early childhood.CLE is almost always unilateral with a male preponderance (M: F=3:1). This disease is characterized by hyperinflation of one or more lobes of the lung, leading to compression atelectasis on the ipsilateral or contralateral side and mediastinal shift. These, in turn, produce ventilationperfusion mismatch. [1] We report this rare case from a northern state of India with the peculiarity of being found in an adolescent with bilateral involvement who is on conservative treatment.

A 13-years-old male presented to our hospital with Breathing difficulty with past H/0 similar episodes since early childhood, denying any history of fever, hemoptysis. On general physical examination, pulse of 100 beats per minute, blood pressure of 110/70 mmHg and respiratory rate of 38 per minute were recorded. Examination of chest revealed a hyperinflated chest. Percussion note over the bulged area was hyper-resonant [Figure 1]. Tactile vocal fremitus and breath sounds were decreased in intensity over bilateral infrascapular & inframammary area. Cardiac and abdominal examination was normal. Immediately X-ray of chest was done, which showed trachea shifted towards right, Emphysematous chest with fibrotic lesions in the midzone, alveolar shadowing present, ? pan acinar emphysema [Figure 2]. Arterial blood gas analysis of the patient showed pO2 -86 mmHg, saturation -94%, pCO2 -42 mmHg and pH -7.39. Serum potassium was 4.1 meq/l, sodium 136 meq/l and bicarbonate 25 meq/l. Electrocardiography and blood chemistry was normal.CT of chest shows extensive marked lobular emphysematous changes seen bilaterally with predominant involvement of lower lobes.

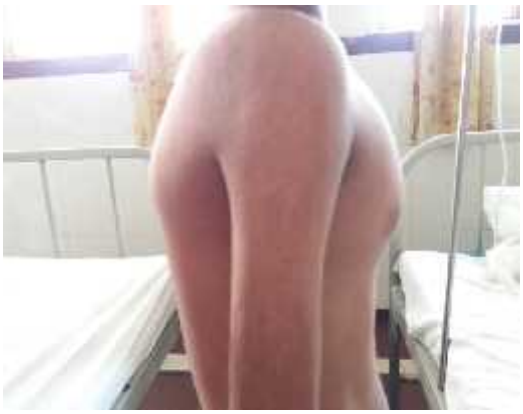

Figure 1

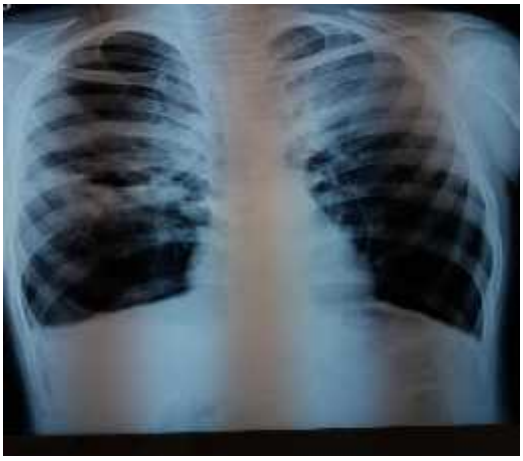

Figure 2

Middle and lingular lobes are also involved. Mild bronchial dilatation is also seen. Pulmonary vessels are markedly attenuated in the involved area. Upper lobes are relatively

spared with few fibrotic bands seen in right upper lobe in peribronchial distribution. Mediastinum is partially compressed. F/S/O Marked Panacinar Emphysema. [Figures 3a and 3b].

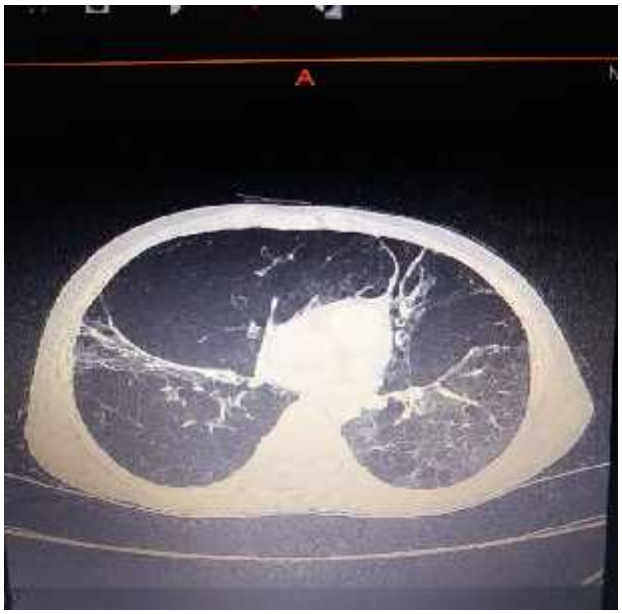

Figure 3a

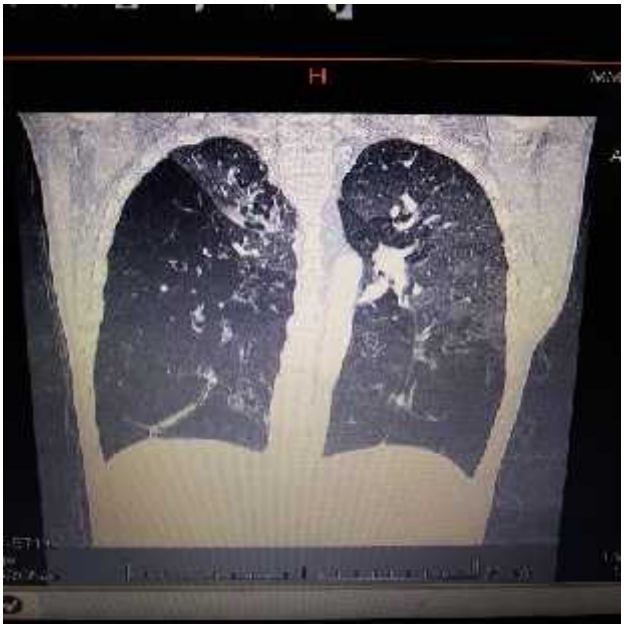

Figure 3b

No bronchocele or intraluminal foreign body was seen and lung parenchyma was normal. In addition, no intrinsic or extrinsic mass lesion of the airways was identified. Pulmonary function tests were FEV1- 86.66% of predicted, FVC-90.60% of predicted, FEF25-75 -77.96% of predicted, VC -2.48L, FEV1/VC-88.3% and MVV -73.6L. Serum 1-antitrypsin level was 121 mg/dl. Lastly, echocardiography was done to find any cardiac lesion that was normal.

DISCUSSION

CLE is characterized by variable severe overinflation of a pulmonary lobe, and compression of surrounding lung parenchyma. Pulmonary vascular markings can be seen up to the periphery of the affected lobe and there is no pleural demarcation as we see in pneumothorax. Mostly single lobe on one side is involved; however, multilobar involvement can be seen. Pulmonary lobes commonly involved are left upperlobe (43%), right middle lobe (32%), and right upper lobe (20%). [1] Adult presentation is unusual. They usually come with exertional dyspnoea or pulmonary infections. Symptoms can be mild and sometimes no symptoms can be seen.

The etiology of CLE is difficult to determine and no apparent cause is found in 50% of cases. Most commonly, the cause is a congenital defect of cartilage, ranging from hypoplastic and flaccid tissue to complete absence, accounting for 25% of cases. The remaining 25% have other causes of bronchial obstruction, such as redundant mucosal fold, mucus plugging, anomalous cardiopulmonary architectures and rarely intrathoracic masses.[2]

There are now data from molecular, genetic and embryonic organ culture studies that indicate that normal primary branching pattern of lung development is regulated by reiterative signaling of a fibroblast growth factor-10 pathway. Secondary branching appears to be dependent on the influence of sonic hedgehog and one of the Homeobox genes, Nkx 2.1, also identified as thyroid transcription factor-1. Although significant mutations in the genetic material controlling these functions appear to result in major anomalies, minor errors in transcription might result in localized deficiencies in the bronchial cartilage leading to the development of CLE. The reiterative nature of signaling also might help in explaining cases in which multiple lobes are affected.[3]

Association between congenital heart disease and CLE has been described. Murray *et al*. showed a 14% incidence of congenital heart diseases among 166 CLE patients. Common cardiac defects were left to right shunt, tetrology of fallot, right sided aortic arch and patent ductus arteriosus.[4,5]

Pulmonary function tests in CLE can be normal in asymptomatic or mild disease, but obstructive pattern is a feature of moderate to severe disease. Radiologically, the cardinal features are overinflation and air trapping, the former is manifested by markedly increased volume of the affected lobe, depressing the ipsilateral diaphragm, compressing the surrounding lung parenchyma and displacing the mediastinum. Vascular markings in the affected lobe are attenuated but maintained till the periphery of the lung. There is no line of pleural demarcation in contrast to pneumothorax. Chest X-ray and CT scan usually establish the diagnosis. Other imaging modalities include MRI and V/Q scan. MRI demonstrates any vascular lesion causing external compression but it is only an adjunct to the diagnosis of CLE, not a primary investigation. V/Q scan depicts ventilation defects and decreased perfusion in the hyoperexpanded lobe due to attenuated vascularity. [6]

Pulmonary lobectomy of involved lobe is the treatment of choice for CLE. [7] PS Critchley and colleagues [8] reported a young pregnant female with left upper lobe CLE and managed by the resection of the involved lobe. [8] Granato F [9] reported first case of endoscopic parenchymal sparing resection in CLE with mild symptoms. Thus video assisted thoracoscopic surgery (VATS) seems to be an emerging and advantageous approach. Dutta *et al*. from India reported conservative treatment of a neonate with CLE and Meir Mei-Zahav *et al*. managed 14 children (0-17 years) conservatively

out of 20 children with CLE. However, studies are further required to determine the long-term outcome of conservative management, as the disease is rare in adults. Going through the literature, surgical option is the suitable one and VATS can be promising in future.

References

1. Kennedy CD, Habibi P, Matthew DJ, Gordon I.Lobar emphysema: Long term imaging follow up. *Radiology* 1991; 180:189-93.
2. Kumar B, Agrawal LD, Sharma SB. Congenital bronchopulmonary malformations: A single-centre experience and a review of literature. *Ann Thorac Med* 2008; 3:135-9.
3. Perl AK, Whitsett JA. Molecular mechanism controlling lung morphogenesis. Clin. *Genet* 1999; 56:14-7.
4. Murray GF, Talbert JL, Haller JA Jr. Obstructive lobar emphysema of new born infant. Documentation of mucous plug syndrome with successful treatment with bronchotomy. *J Thorac Cardiovasc Surg* 1967;53:886-90.
5. Ozcelik U, Gocmen A, Kiper N. CLE: Evaluation and long term follow up of 30 cases at a single centre. *Paediatr Pulmonol* 2003; 35:384-91.
6. Karnak I, Senocak ME, Ciftci AO, Büyükpamukçu N. *et al*. C L E-Diagnostic and therapeutic considerations. *J Paed Surg* 1999; 34:1347-51.
7. Sathish Kumar T, Simon A, Sen S. Bilateral Congenital Emphysema: Unusual cause for recurrent respiratory tract infection. *J Indian Assoc Paed Surg* 2006; 11:151-2.
8. Critchley PS, Forrestr-Wood CP, Ridly PD. Adult Congenital Lobar Emphysema in pregnancy. *Thorax* 1995; 50:909-10.
9. Granato F, Voltolini L, Spina D, Paladini P, Gallazzi M, Gotti G.VATS for congenital lobar emphysema: A case report. *Minerva Chir* 2009; 64:111-5.
10. Dutta AK, Mandal S, Jana J. Congenital lobar emphysema: A case report. *Cases J* 2009; 2-67.
11. Mei-Zahav M, Konen O, Manson D, Langer JC. Is congenital lobar emphysema a surgical disease?. *J Pediatr Surg* 2006; 41:1058-61.

****