Congenital lobar emphysema: Is surgery routinely necessary?

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ABSTRACT

Congenital lobar emphysema (CLE) is a rare congenital abnormality characterised by overinflation of a pulmonary lobe. Its aetiology is unknown. The management of CLE has traditionally been surgical. A newborn boy with a birthweight of 2.5 kg was delivered at full-term by caesarian section due to food delivery. There was no marked respiratory distress at birth, and little meconium stained liquor was seen on the skin. The initial diagnosis was meconium aspiration syndrome. After computed tomography of the thorax, CLE was diagnosed. The patient was observed throughout for a week and the CT of the thorax was repeated, which revealed that the emphysema had resolved. The nonoperative approach should be considered in asymptomatic patients with CLE.

Key words: Rare lung disease, thoracic surgery

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INTRODUCTION

Congenital lobar emphysema (CLE) is a rare congenital abnormality characterised by overinflation of a pulmonary lobe.[1] Its aetiology is unknown. Clinical presentation varies from acute neonatal respiratory failure to recurrent episodes of tachypnoea or infections.[2,3] The management of CLE has traditionally been surgical. Because of the increased use of imaging, this lesion is frequently found in asymptomatic and mildly symptomatic children, prompting us to adopt a more conservative approach to these children.[4] This report describes a neonate with asymptomatic CLE; and the CLE was diagnosed at the age of 2 days in this child.

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CASE REPORT

A newborn boy with a birthweight of 2.5 kg was delivered at full-term by caesarian section due to food delivery. There was no marked respiratory distress at birth and little meconium stained liquor was seen on the skin. So, the patient was admitted to the newborn unit. On the physical examination, there was no intercostal retraction, cyanosis and tachypnoea. Breath sounds were diminished on the right side. The blood parameters were normal.

The initial chest x-ray, taken shortly after birth, showed hyperinflation on the right side. CT of the thorax supported the plain x-ray. There was hyperinflation in the right middle lobe and the anterior segment of the left upper lobe on the left side [Figure 1].

The patient was observed throughout for a week and then a CT scan of the thorax was done. The emphysema was resolved [Figure 2].

DISCUSSION

CLE is a rare cause of respiratory distress during infancy which is cured by surgery. CLE has a prevalence of approximately 1:20,000 to 1:30,000.[3] In 25% of the cases, this developmental anomaly is attributed to a congenital cartilage defect,[5] whereas in another 25%, the development of CLE is caused by bronchial obstruction.[5,6] In 50% of the cases, a clear aetiology cannot be identified.[6] It is usually unilateral, affecting the left upper lobe, followed by the right middle lobe, although bilateral involvement has been reported.[7,8] Most cases of CLE become symptomatic; respiratory distress in the neonatal period may be life threatening. In as many as 25% of the cases, presentation is delayed until the first month of life.[9]

In this case report, the neonate presented with asymptomatic CLE. In keeping with the clinical scenario and physical examination, the initial diagnosis was mistaken for meconium aspiration syndrome. A

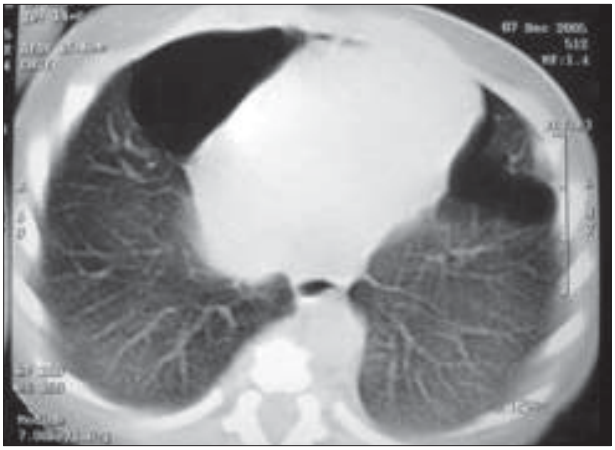


Figure 1: Axial computed tomography shows hyperinflation in both lungs

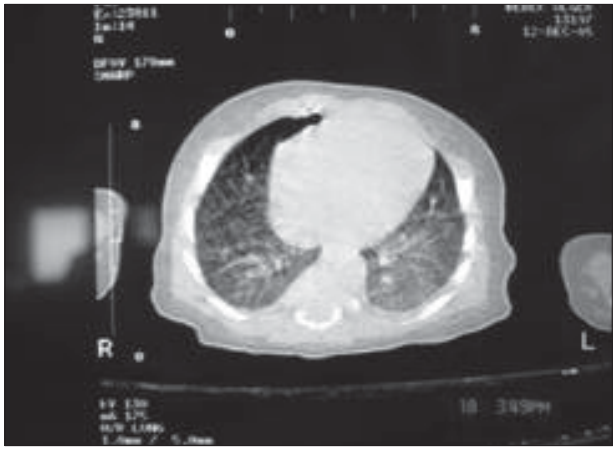


Figure 2: After one week emphysema resolved almost completely

chest x-ray and CT scan were necessary to refute this diagnosis. CT is an important diagnostic procedure in CLE, because it can exclude a vascular abnormality and other conditions that might be confused with CLE such as pneumothorax, pneumatocoele, diaphragmatic herniation, or cystic adenomatoid malformation.[10]

Although, it seems logical to recommend surgery for severely symptomatic patients, and a nonoperative approach for asymptomatic lesions, the approach to children with mild symptoms is less clear. It should be appreciated that the asymptomatic or mildly symptomatic children with CLE can deteriorate and present with life threatening events later in life[4]. Tharkal *et al.* described seven children with mild symptoms who were initially managed conservatively. Out of these seven

four had to undergo surgery ultimately within the following few months.[3] Other studies describe a small proportion of patients treated conservatively with a good outcome.[11,12]

Conservative and surgical treatments have similar outcomes after a long term follow-up.[13] CLE usually follows a progressive course and if, not recognised and treated by surgery, it may be fatal. However, the nonoperative approach should be considered in asymptomatic patients with CLE.

REFERENCES

1. Chun CC, Huang SC, Liu MC, Se TY. Fetal congenital lobar emphysema. Taiwanese J Obstet Gynecol 2007;46:73-6.
2. Schwartz MZ, Ramachandran P. Congenital malformations of the lung and mediastinum—a quarter century of experience from a single institution. J Pediatr Surg 1997;32:44-7.
3. Thakral CL, Maji DC, Sajwani MJ. Congenital lobar emphysema: experience with 21 cases. Pediatr Surg Int 2001;17:88-91.
4. Mei-Zahav M, Konen O, Manson D, Langer JC. Is congenital lobar emphysema a surgical disease? J Pediatr Surg 2006;41:1058–61.
5. Pardes JG, Auh YH, Blomquist K, Kazam E, Macid M. CT Diagnosis of congenital lobar emphysema. J Comput Assist Tomogr 1983;7:1095-7.
6. Kravitz RM. Congenital malformations of the lung. Pediatr Clin North Am 1994;41:453-72.
7. Miller CG, Woo-Ming MO, Carpenter RA. Lobar emphysema of infancy. Case report of bilateral involvement with congenital heart disease. West Indian Med J 1968;17:35-40.
8. May RL, Meese EH, Times JJ. Congenital lobar emphysema: case report of bilateral involvement. J Thorac Cardiovasc Surg 1964;48:850-4.
9. Armstrong P, Wilson A G, Dee P, Hansell DM. Congenital lobar emphysema. In: Armstorng P, Wilson AG, Dee P, Hansell DM, editors. Imaging of diseases of the chest. London: Mosby; 2000 p. 695-7.
10. Stanton M, Davenport M. Management of congenital lung lesions. Early Human Development 2006;82:289-95.
11. Al-Bassam A, Al-Rabeeah A, Al-Nassar S, Al-Mobaireek K, Al-Rawaf A, Banjer H, *et al*. Congenital cystic disease of the lung in infants and children (experience with 57 cases). Eur J Pediatr Surg 1999;9:364-8.
12. Karnak I, Senocak ME, Buyukpamukcu N. Congenital lobar emphysema: diagnostic and therapeutic considerations. J Pediatr Surg 1999;34:1347-51.
13. Eigen H, Lemen RJ, Waring WW. Congenital lobar emphysema: long term evaluation of surgically and conservatively treated children. Am Rev Respir Dis 1976;113:823-31.

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