

Bilateral congenital emphysema: Unusual cause for recurrent respiratory tract infection

T. Sathish Kumar, Anna Simon, Sudipta Sen*

Departments of Child Health and *Pediatric Surgery, Christian Medical College and Hospital, Vellore, India

Correspondence: Dr. Anna Simon, Child Health Unit I, Christian Medical College, Ida Scudder Road, Vellore - 632 004, India.
E-mail: child1@cmcvellore.ac.in

ABSTRACT

Congenital lobar emphysema is usually restricted to one lobe of a lung. We report a rare case of bilateral congenital lobar emphysema, where the right middle lobe and its equivalent on the left side (lingula) were affected. Newer modalities of imaging (like high resolution CT scan thorax) have made the simultaneous diagnosis of bilateral involvement possible, which otherwise could be missed in a plain chest radiograph.

KEY WORDS: Bilateral congenital lobar emphysema, recurrent respiratory tract infection

CASE REPORT

A 6-month-old male infant presented with a history of recurrent respiratory infections with failure to thrive since infancy. There was associated cough, fast breathing and lower chest in-drawing since the age of 1 month. He needed hospitalization often and treatment with IV fluids and parenteral antibiotics for each episode. There was no history of edema, loose stools or recurrent skin or ear infections.

On examination, his weight was 4 kg, and he was pale. He had no cyanosis, clubbing or generalized lymphadenopathy. He was tachypnoeic, and his saturation was 88% on 5 L of oxygen. His respiratory system examination revealed a trachea in normal position with hyper-inflated chest and hyper-resonant percussion note bilaterally. He had decreased intensity of vesicular breath sounds in both infraclavicular and mammary areas with crepitations all over the chest. Rest of the systemic examination parameters were within normal limits.

Clinical diagnosis for evaluation considered at admission was recurrent pneumonia with failure to thrive. Chest radiograph revealed bilateral hyper-inflated lung fields [Figure 1]. His hemoglobin was 12 gm%, total count 12,900 cells/cu. mm, differential count of BF2%, N59%, L39% and platelet count 391,000/cu. mm. His serum sodium was 137 mEq/L, potassium was 3.4 mEq/L and α -1 antitrypsin level was 154 U/L. His immunoglobulins were normal for age, and sweat chloride was 32 mEq/L. Barium swallow did not reveal any gastroesophageal reflux or H-shaped tracheo-esophageal fistula. The



Figure 1: Chest radiograph showing bilateral hyperinflated lung fields

echocardiogram was normal. Mantoux test and gastric juice for AFB were negative. He was given parenteral antibiotics. Nutritional support was given as nasogastric feeds supplemented with trace elements and multivitamins.

A high-resolution CT scan thorax was done, which revealed emphysematous changes in the right middle lobe and left lingula with atelectatic changes in adjacent lobes [Figure 2]. Since the child continued to deteriorate with medical management alone, surgical intervention was considered. A left thoracotomy was done initially, and per-operatively the left lingular segment was found to be emphysematous with collapse of the left upper and lower lobes. On removal of the lingula, there was good expansion

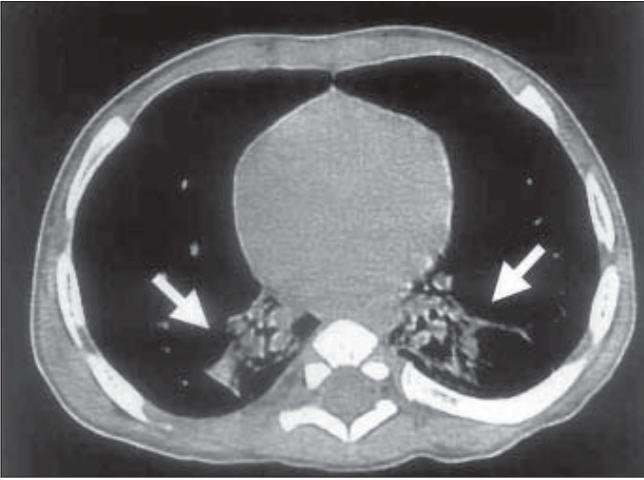


Figure 2: CT scan thorax showing emphysematous right middle and lingula with collapsed lower lobes

of the left upper and lower lobes. On the right side, the right middle lobe was found to be emphysematous with collapse of the right upper and lower lobes. The collapsed segments expanded after the right middle lobe was excised. Histopathological examination of excised emphysematous lobes revealed features consistent with congenital lobar emphysema of both the lingula and the right middle lobe. The sections of bronchi in the hilar region revealed absence of cartilage in their walls.

In the immediate postoperative period, his oxygen requirement reduced from 5 L/min (preoperative period) to 2 L/min to maintain oxygen saturation of 94%. After 14 days of postoperative period, he worsened and required increasing concentrations of oxygen to maintain normal arterial oxygen concentration. He continued to gradually worsen and expired a month after surgery. Postmortem lung biopsy did not disclose any abnormality in the remaining lung.

DISCUSSION

Congenital lobar emphysema (CLE, Pan lobular emphysema of infancy) is the postnatal over-distension of one or more lobes of a histologically normal lung, usually presenting with respiratory distress in infancy.^[1]

Congenital lobar emphysema is characterized by progressive over-distension due to air trapping in the affected lobe, concomitant compression of the remaining lung tissue, displacement of mediastinum with herniation of lung to the unaffected side and flattening of diaphragm on the affected side.^[2] In the case under report, as the involvement was bilateral, the classical radiological manifestations of congenital lobar emphysema mentioned above were absent.

Etiological mechanism in CLE is still obscure. An

obstructive abnormality of the bronchus seems to be the most obvious explanation for the development of CLE. Various intrinsic bronchial abnormalities, including abnormal mucus folds, bronchial stenosis, bronchial kinking or rotation, have been found in patients with CLE. Bronchomalacia is the most common abnormality. It can be focal or diffuse. The abnormal cartilage can be absent, hypoplastic, flaccid or immature.^[3] Histopathological examination in this child revealed absence of bronchial cartilage in the emphysematous lobes which were removed.

Plain chest radiograph in CLE is often diagnostic and shows findings typical of lobar hyperinflation. Serial chest X rays show increasing hyperinflation of the emphysematous lobe with progressive atelectasis of ipsilateral normal lung, mediastinal shift away from the emphysematous side and compression of contralateral lung. The upper and middle lobes are more involved than the lower lobes, and the left lung is more often involved than the right.^[3] In most cases, only a single lobe is involved, but two or more emphysematous lobes have been noted.^[2,4,5]

There were only a few case reports in the literature about bilateral congenital lobar emphysema which were surgically treated at one sitting^[2] or treated one side at a time followed by the other.^[4,5] With the advent of newer modalities of imaging like high resolution CT scan of the thorax and MRI, the simultaneous diagnosis of bilateral involvement is made possible as in our case and as in a case reported in 1987.^[2] The diagnosis of bilateral involvement in the other reports^[4,5] was made sequentially, the classical manifestations of CLE becoming evident on one side after removal of the affected emphysematous lung on the opposite side. Surgical correction of bilateral congenital lobar emphysema was done in two stages in the other two reported cases.^[4,5] Simultaneous diagnosis of bilateral CLE has made possible successful surgical treatment of bilateral lobar emphysema at one operative session.^[2]

REFERENCES

1. Biswal N, Mathai R, Bhatia RD, Bhat BV, Puri RK, Karthikeyan G, *et al.* Congenital lobar emphysema. *Indian Pediatr* 1993;30:1349-54.
2. Ekkelkamp S, Vos A. Successful surgical treatment of a newborn with bilateral congenital lobar emphysema. *J Pediatr Surg* 1987;22:1001-2.
3. Lierl M. Congenital abnormalities. In: *Pediatric Respiratory Disease*. Hilman BC (editor). W.B. Saunders: Philadelphia; 1993. p. 468.
4. May RL, Meese EH, Timmes JJ. Congenital lobar emphysema: Case report of bilateral involvement. *J Thorac Cardiovasc Surg* 1964;48:850-4.
5. Floyd FW, Repici AJ, Gibson ET, McGeorge CK. Bilateral congenital lobar emphysema surgically corrected. *Pediatrics* 1963;31:87-96.