CASE REPORT

Mediastinal bronchogenic cyst mimicking congenital lobar emphysema

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SUMMARY

Bronchogenic cyst (BC) is a rare congenital malformation of the lung. Most patients remain asymptomatic until adulthood while some are symptomatic in the first few years of life. However, symptoms in newborn period are rare. We report a case of a 3-day-old preterm baby with respiratory distress diagnosed as congenital lobar emphysema on chest X-ray. A CT scan revealed a mediastinal cyst causing obstructive lobar emphysema. The cyst was excised and pathological examination was suggestive of BC.

BACKGROUND

Bronchogenic cyst (BC) occurs as a developmental anomaly of the primitive foregut and can be mediastinal or pulmonary in location. It may be asymptomatic, but most cases become symptomatic in the second or third decade of life.

There have been very few documented cases of BC presenting as respiratory distress in neonatal period. We present a rare case of a 3-day-old newborn with a mediastinal BC who presented with unilateral hyperinflation of the lung mimicking congenital lobar emphysema (CLE) on chest X-ray.

CASE PRESENTATION

A 1.2 kg baby girl was born at 34 weeks by lower segment Caesarean section for severe pre-eclampsia in mother with an Apgar of 9 and 10 at 1 and 5 min, respectively. The baby had normal respiration and was saturating well on room air. The mother had regular antenatal care and a morphological scan was normal.

On the third day of life, she developed respiratory distress in the form of tachypnoea with chest retractions. Capillary refill time was normal. Examination revealed a respiratory rate of 78/min, heart rate of 148 bpm and blood pressure of 64/36 mm Hg. Oxygen saturation decreased to 84% on room air. Heart sounds were normal. Anterior fontanelle was normal. Breath sounds were decreased on the left side.

INVESTIGATIONS

Blood counts were normal, blood gas showed pH 7.18, pCO₂ 37.5 mm Hg, HCO₃ 13.6 mmol/L and base excess -13.5 mmol/L. Chest X-ray revealed hyperinflation of left side with mediastinal shift to right (figure 1). Pulmonary vascular markings were seen on the left side. Transillumination test was negative for pneumothorax, and echocardiogram and neurosonogram were normal.

DIFFERENTIAL DIAGNOSIS

A preterm baby developed tachypnoea and metabolic acidosis on third day of life. Differential diagnoses considered were neonatal sepsis, pneumonia, patent ductus arteriosus and intraventricular haemorrhage.

Chest X-ray showed hyperinflation of left lung with mediastinal shift to opposite side. The possibility of CLE and pneumothorax was considered. Pneumothorax was ruled out as transillumination was negative and bronchovascular markings were seen on the left side.

A CT thorax performed on day 8 of life for persistent tachypnoea showed a cyst in the mediastinum between the trachea and oesophagus causing narrowing of the left main bronchus and hyperinflation of the left lung (figure 2). The differential diagnosis based on CT thorax findings was BC and oesophageal duplication cyst.

TREATMENT

In view of suspected sepsis, the baby was started on antibiotics. She was administered oxygen for desaturation via nasal catheter at 2 L/min. One normal saline bolus was administered and dobutamine was started for metabolic acidosis. Her general condition improved, metabolic acidosis resolved, blood pressures remained normal and oxygen was discontinued as saturation normalised on room air. However, the baby continued to be tachypnoeic. CLE was suspected based on chest X-ray. Blood culture showed no growth and sepsis screen was negative. Therefore, antibiotics were stopped after 5 days.

Figure 1 Frontal radiograph of chest showing hyperinflation of left lung with trans-herniation of left lung to right side along with mediastinal shift.

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Figure 2 Axial and coronal sections of thorax CT showing a well-defined cystic lesion in middle mediastinum between trachea and oesophagus in (A), with inferior part of the lesion compressing left main bronchus in (B and D) resulting in hyperinflation of entire left lung, trans-herniating to right side in (C) with contralateral mediastinal shift.

In view of persistent tachypnoea, CT thorax was performed on day 8, which showed mediastinal cyst causing obstructive lobar emphysema. Conservative management was planned, as the baby was preterm, weighed 1.2 kg and was clinically stable on room air. Respiratory distress worsened on day 24 of life. Examination showed respiratory rate of 82/min with severe chest retractions. The baby was started on antibiotics and surgery for mediastinal cyst was planned. The baby underwent left thoracotomy. A 2×2 cm bluish cyst was seen in the posterior mediastinum between the trachea and oesophagus. The arch of aorta was seen to wrap the cyst superiorly and laterally. The cyst was excised and pathological examination showed that the cyst was lined by respiratory epithelium (figure 3). The wall contained seromucinous glands, smooth muscle cells and hyaline cartilage (figure 4). These findings were suggestive of BC. The baby was ventilated for 4 days postsurgery and extubated to room air after which she remained asymptomatic. Antibiotics were given for 7 days.

OUTCOME AND FOLLOW-UP

The baby was seen in follow-up outpatient department at 6 months of life. She was active, thriving well and without respiratory distress.

DISCUSSION

BCs are congenital cystic malformations of the lungs, which arise as abnormal buds from ventral portion of the embryonic

Figure 3 Photomicrograph showing cyst lined by respiratory epithelium (H&E ×100 magnification).

foregut. They do not branch further and end as blind pouches. They can be mediastinal (75%) or intrapulmonary (20%). BCs are usually asymptomatic in the first few years of life and can present at any age.¹ 50% of cases are asymptomatic until adulthood.² BCs are lined by respiratory epithelium and the walls contain smooth muscle and hyaline cartilage. They may or may not communicate with the tracheobronchial tree.

Figure 4 Photomicrograph showing cyst containing seromucinous glands (H&E ×100 magnification).

BCs are known to produce compressive symptoms on the tracheobronchial tree and cardiac structures. There has been few case reports of BC masquerading as CLE in infants and children.^{3–5} In this case, the baby was symptomatic from third day of life and a diagnosis of CLE was made based on chest X-ray. A CT scan showed a mediastinal cyst later found to be BC after pathological examination, compressing left main bronchus leading to obstructive lobar emphysema, a rare presentation in neonates. We also note that this lesion was not diagnosed antenatally, but BCs are known to progress or regress with time. BCs can also present as tracheal stenosis or oesophageal compression without evidence of lung overinflation and CT thorax will aid in the diagnosis.⁶

Oesophageal duplication cyst can arise in this location from posterior division of primitive foregut and is lined by squamous epithelium. Pathological examination helped to differentiate it from BC as in this case the cyst was lined by respiratory epithelium.

Idiopathic CLE commonly affects left upper lobe. Radiologically there will be evidence of collapse of neighbouring lobes. Multiple lobes may be involved in 5% of cases only.^{7,8} In our case, the hyperinflation of the entire left lung points against CLE.

Obstructive lobar emphysema may be due to intrinsic or extrinsic causes. These constitute less than one-third of all CLE. Intrinsic causes are defective cartilage, bronchial mucosal folds and webs. Here we focus on obstructive lobar emphysema caused by extrinsic compression. Causes are BC, tumours, vascular causes, lymph nodes and cardiac causes. A CT scan is sensitive in most cases to delineate the lesion. In cases where

the main bronchus is compressed, the entire lung becomes hyperinflated.

The management in obstructive lobar emphysema secondary to BC involves surgical excision of cysts unlike CLE where lobectomy will be needed. Most cases of BCs become symptomatic, hence surgical excision is recommended even in asymptomatic patients.^{9,10}

Learning points

- ▶ In all cases of suspected congenital lobar emphysema (CLE), obstructive causes must be ruled out.
- ▶ Radiologically CLE commonly involves left upper lobe with compression of adjacent lung while obstructive lobar emphysema due to compression of main bronchus causes hyperinflation of whole lung.
- ▶ A routine CT scan in all cases of suspected CLE may be helpful to rule out obstructive causes.

Contributors All the doctors who were involved in the diagnosis and management of the baby. SA and BJR collected data and wrote the paper. MA helped in literature review and reviewed the manuscript.

Competing interests None declared.

Patient consent Obtained.

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