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SHORT COMMUNICATION

Bronchogenic cyst: An uncommon cause of congenital lobar emphysema

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KEYWORDSBronchogenic cyst;
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Dyspnoea**Summary**

We report a case of a 1-month-old boy who has developed respiratory distress. Chest X-ray and CT scan showed over distension of the left upper lobe and a mediastinal shift in favour of congenital lobar emphysema (CLE) of the left upper lobe. One month after uneventful lobectomy, he was readmitted at hospital for another episode of respiratory distress. Chest radiography revealed relapse of compressive emphysema in the remaining left lobe. Gastro oesophageal transit and MRI were performed, which have shown a mediastinal cystic mass. Accordingly, the patient underwent thoracotomy. Surgical examination found a subcarinal bronchogenic cyst which compressed the main left bronchus, causing the CLE of both upper and lower left lobes. Histological examination of removed cyst confirmed these data. Authors discuss causes of diagnostic delay.

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Introduction

Bronchogenic cyst (BC) is an unusual congenital lung development anomaly, rarely reported in infants.¹ It may lead to various symptoms and complications. Mediastinal BC in children is often responsible for compressive symptoms of tracheo-bronchial tree because of its relative softness.^{2,3} We report here, a case of an infant with BC revealed by congenital lobar emphysema (CLE) and in

which, BC was misdiagnosed at first time. We discuss causes of diagnostic delay.

Case report

A 1-month-old baby boy was admitted to Children's Hospital of Tunis with respiratory distress. He was born after a normal pregnancy of 40 weeks. His birth weight was 4200 g. He was exclusively breast-fed and had no history of family diseases. Three days before, he developed cough and respiratory embarrassment. His respiratory status worsened so much, that he was hospitalised. Physical examination showed marked dyspnoea, cyanosis and no rales at pulmonary auscultation. He had neither

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Figure 1 Chest radiography: note patulous left lung, mediastinum deviation and right upper lobe atelectasis.

fever nor clinical symptoms of heart failure. Gasometry showed a hypoxia at 63.8 mmHg, a capillary oxygen saturation at 93% and a capnia at 48.7 mmHg. Chest radiography showed over distension of left lung, a mediastinum deviation and a right upper lobe atelectasis. (Fig. 1). Chest CT scan was in favour of an upper left lobe emphysema (Fig. 2). Lobectomy was then performed without any complication. Patient was discharged within 10 days. Microscopic examination revealed an alveolar distension and atrophic inter-alveoli septum. One month later, he was readmitted with 3-days history of fever, cough and marked dyspnoea. Physical examination showed nasal hyrorrhoea, fever and marked wheeze. Bronchiolitis was then suspected. Chest X-ray revealed relapse of compressive emphysema in the remaining left lobe (Fig. 3). Complementary radiological investigations were performed to look for an eventual compressive-mediastinal anomaly.



Figure 2 Chest CT scan: upper left lobe emphysema.



Figure 3 Chest radiography: compressive emphysema in the remaining left lobe.

Gastro oesophageal transit showed an extrinsic regular print in the oesophagus anterior side (Fig. 4). MRI confirmed the emphysema of the left lower lobe (LLL) and identified a cystic mass in the posterior and medium mediastinum, between trachea and oesophagus (Fig. 5). These data were in favour of bronchogenic cyst compressing left main bronchus and causing CLE. So, patient underwent thoracotomy. A subcarinal bronchogenic cyst was so adherent to trachea and main left bronchus that resection created a small defect in the tracheal membranous wall. This defect was closed successfully without further complications. A subtotal resection of cyst was performed without touching the emphysematous lobe. The removed cyst measured 3 cm and was filled with a mucoïd content. Microscopic examination showed typical features with



Figure 4 Gastro oesophageal transit: note extrinsic regular print in the oesophagus anterior side.

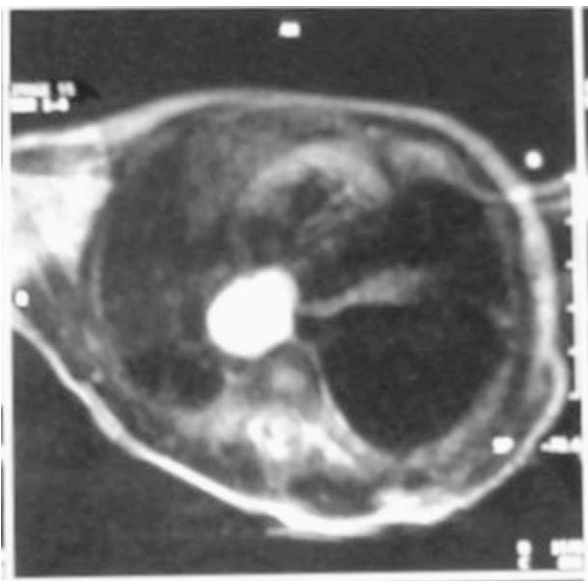


Figure 5 Chest MRI: note lower left lobe emphysema and cystic mass in the posterior and medium mediastinum, between trachea and oesophagus.

ciliated columnar-epithelial lining and connective tissue wall with cartilage. Post-operative course was characterised by relapse of respiratory embarrassment attacks within early months. One year later, he became a symptomatic boy. Left lower lobe looked less emphysematous on subsequent Chest X-ray (Fig. 6).

Discussion

Both congenital lobar emphysema (CLE) and bronchogenic cyst (BC) are rare anomalies of lung development. CLE is



Figure 6 Chest radiography: left lower lobe looks less emphysematous.

characterised by over inflation of pulmonary lobe due to air trapping by a check-valve mechanism.⁴ BC arise from abnormal budding of the primitive tracheo-bronchial tree during airway development.^{5,6} Typically, CLE is responsible for progressive respiratory distress that occurs during the first few weeks of life.^{4,7,8} BC can lead to various symptoms and complications that often occur later, in children aged more than 6 years or even in adults.^{1,2,9} Congenital heart diseases are often reported with CLE and they are sometimes involved in the bronchial extrinsic obstruction mechanism.^{10,11} However, congenital lobar emphysema due to a bronchogenic cyst has been rarely reported.¹²

Bronchogenic cyst was misdiagnosed at the beginning by clinicians, radiologists and surgeons. If patient respiratory status had allowed performing bronchoscopy before surgery, bronchial compressive process could have suspected before. It's also possible that the BC was too small to be detect by CT scans. It's clear that surgical exploration of the deep mediastinum was not exhaustive in the first operation. Finally, we think that lobectomy offered possibility for the cyst to increase in its volume and then to become easily recognisable by imaging process.

Conclusion

Although BC is an uncommon cause of CLE, clinicians should look for as possible mechanism involved in extrinsic bronchial compression. In addition to careful radiographic analysis, bronchoscopy when possible, may be useful for aetiological diagnosis. The latter, can be confirmed by exhaustive chirurgical investigation.

Conflict of interest

I also certify that authors have no conflicts of interest including financial or personal relationships.

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