

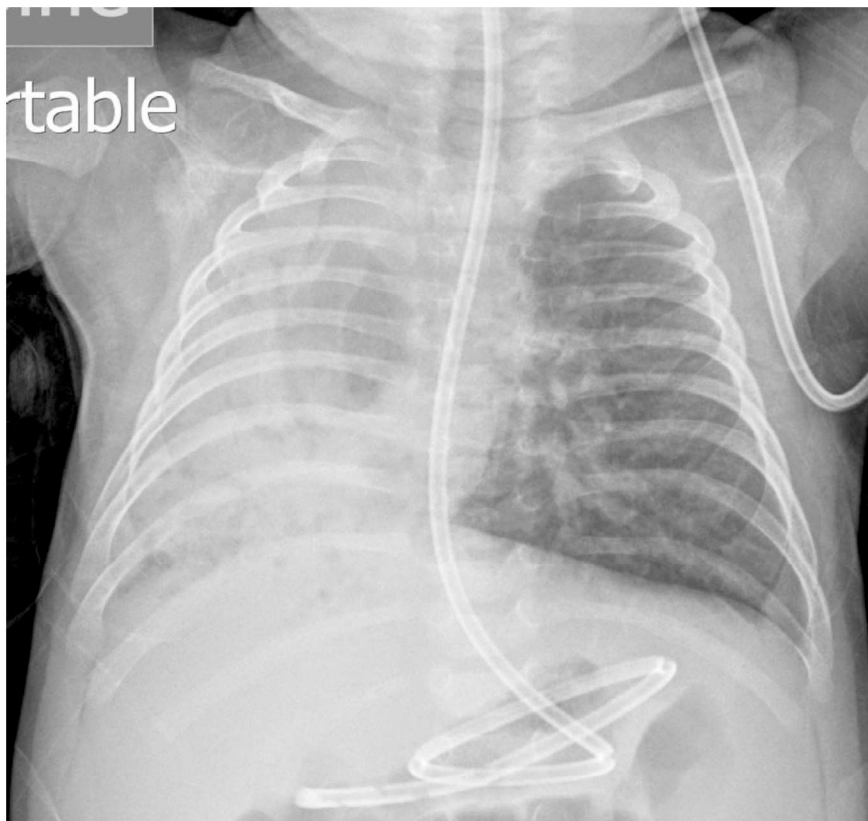


Imaging Quiz

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This is a 2-month-old baby girl with mild respiratory distress in room air. She was born preterm at 34 weeks and was small for gestational age. Her cardiac abnormality was noted in her antenatal scan. She had history of desaturation during oral feeding during her stay in special care baby unit. Nevertheless she tolerated feeding subsequently and had satisfactory weight gain.



Question

What are the chest X-ray abnormalities?

(Answer on page 20)



X-ray Quiz

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This is a 28-day-old baby boy with noisy breathing and stuffy nose for one week before admission. He was born at term with uneventful antenatal and postnatal course. There was no associated fever, cough or nasal discharge. Physical examination reviewed no respiratory distress with a clear chest on auscultation and normal oxygen saturation in room air. Chest radiograph were taken at 1 month (Figure 1a) and 2 months (Figure 1b) of age.

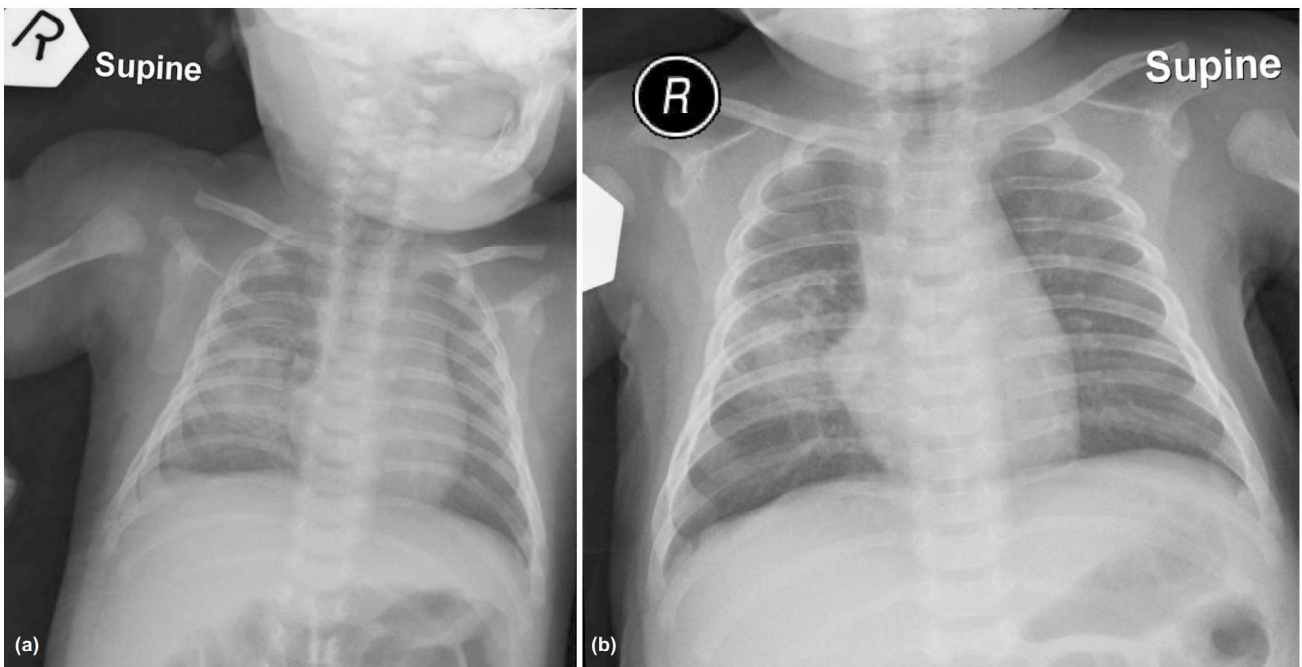


Figure 1. (a) CXR taken at first month of life; (b) CXR taken at second month of life.

Questions

1. What is the abnormality noted on the chest radiographs (CXR)?
2. What further imaging study would be useful?
3. What are the differential diagnoses?

(Answer on page 21)



Answers to Imaging Quiz on page 17

What are the chest X-ray abnormalities?

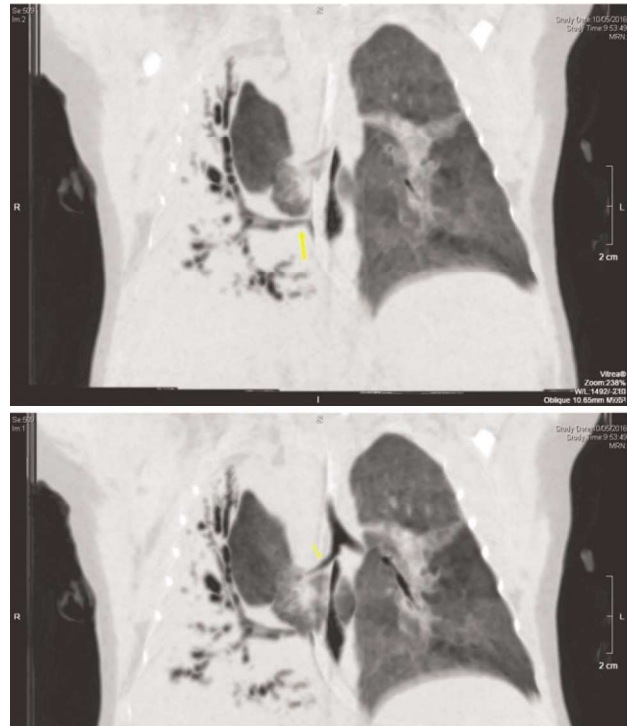
- Right lung opacities and linear lucencies suggestive of consolidation-collapse of right middle and right lower lobes with air bronchogram.
- A small segment of aerated lung in the right upper lobe
- Flattening of left hemi-diaphragm and mediastinal shift to the right suggestive of volume loss in the right lung and compensatory hyper-expansion of left lung
- Dextrocardia
- Transpyloric feeding tube in-situ

Her echocardiogram revealed dextrocardia and hypoplastic right pulmonary artery.

Computed tomography of the thorax was ordered for assessment of structural abnormality in view of persistent right lung shadow. The computed tomography of thorax revealed a bronchoesophageal fistula connecting the distal oesophagus with the right bronchus intermedius which did not communicate with the trachea, consolidation and parenchymal distortion in the right lung and distal tracheal stenosis; sequestered lung was suspected but the vascular supply could not be well demonstrated.

She underwent surgical repair of bronchoesophageal fistula at 4 months old when she achieved target body weight by transpyloric enteral nutrition. The abnormal lung lobes were adherent and non-pliable from recurrent infection and because of dextrocardia, patient had quite severe haemodynamic instability. As result, only a partial lung resection could be performed. Post-operation there was a tiny leakage at the fistula which spontaneously sealed up after 2 months.

Bronchoesophageal fistula is a rare type of anomaly of lung development termed communicating bronchopulmonary foregut malformations. It was thought to be the result of a focal defect in the mesoderm between the lung bud and esophagus during embryological development, and the subsequent rapid growth of esophagus causes separation of the affected lung segment with the trachea.¹ Respiratory distress, chronic cough, choking or desaturation exacerbated by feeding are suggestive clinical features, and recurrent pneumonia is typical. Late presentation in adulthood is also not uncommon.² Diagnosis is confirmed by imaging study. Plain radiograph followed by contrast swallow study to demonstrate the fistula could be sufficient. Computed tomography or magnetic resonance imaging is an alternative which allows multi-planar image reconstruction of the fistula and concomitant assessment of associated parenchymal, airway, and vascular abnormalities. Surgery with resection of the malformed lung segment and division of the fistula is the definitive treatment. Thoracoscopic approach to surgery has been reported with success.^{3,4}



References

1. Srikanth MS, Ford EG, Stanley P, Mahour GH. Communicating bronchopulmonary foregut malformations: classification and embryogenesis. *J Pediatr Surg* 1992;27(6):732-6.
2. Wang CR, Tiu CM, Chou YH, Chang T. Congenital bronchoesophageal fistula in childhood. Case report and review of the literature. *Pediatr Radiol* 1993;23(2):158-9.
3. Katz R, Pitt R, Kim D, Wingrove B. Thoracoscopic pneumonectomy for communicating bronchopulmonary foregut malformation in a 4-month-old child. *J Pediatr Surg* 2010;45(2):427-9.
4. Park T, Jung K, Kim HY, Jung SE, Park KW. Thoracoscopic management of a communicating bronchopulmonary foregut malformation in a 23-month-old child. *J Pediatr Surg* 2012;47(3):e21-3.

Answers to X-ray Quiz on page 18

1. Ill-defined air space opacity is persistently noted at right middle zone on CXR. Associated lucencies are noted within the opacity, could be cystic changes.
2. CT scan of the thorax with IV contrast.
3. Main differential diagnoses include congenital pulmonary airway malformation, pulmonary sequestration, and necrotising pneumonia.

Discussion

CT thorax (Figure 2) was performed in view of persistent abnormality on CXR. Congenital pulmonary airway malformations (CPAM) are multicystic masses of segmental lung tissue with abnormal bronchial proliferation. Patients usually present antenatally on antenatal ultrasound or present during neonatal period with worsening respiratory distress.

Lesions usually involve a single lobe. They less frequently affect the middle lobe, otherwise there is no well-documented lobar predilection.

There are 5 types of CPAM, classified according to the cyst size.

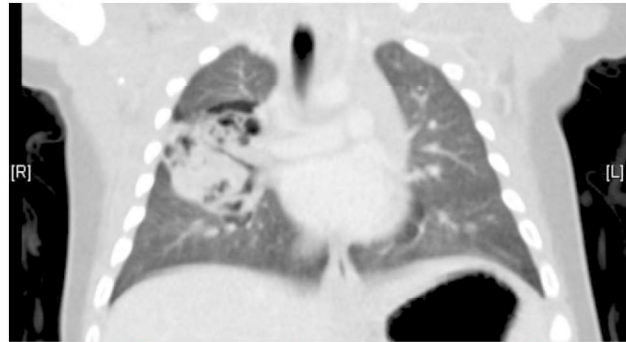


Figure 2. CT thorax shows a heterogeneous mass with internal small cystic components in right upper lobe, with probably involvement of right middle lobe. No definite arterial supply from the aorta is noted. Features suggest congenital pulmonary airway malformation (CPAM). Pathology of resected right lung later shows type III CPAM.

Type	Frequency	Features
Type I	Most common: 70% of cases ¹	<ul style="list-style-type: none"> • large cysts • one or more dominant cysts: 2-10 cm in size • cysts are <2 cm in diameter • associated with other abnormalities
Type II	15-20% of cases ¹	<ul style="list-style-type: none"> • microcysts: <5 mm in diameter • typically involves an entire lobe • has a poorer prognosis
Type III	~10% of cases	<ul style="list-style-type: none"> • unlined cyst • typically affects a single lobe • indistinguishable from type I on imaging²
Type IV	Rare	<ul style="list-style-type: none"> • represents global arrest of lung development³
Type 0	Very rare, lethal postnatally	

Chest radiographs in type I and II CPAMs may demonstrate a multicystic (air-filled) lesion. If seen at the lower lobes, the main differential diagnosis is congenital diaphragmatic hernia. Unlike in diaphragmatic hernia, the distribution of abdominal bowel gas is normal. Air-fluid levels may be seen with or without superimposed infection.⁴ Type III lesions appear solid.

CT is crucial for the management of CPAMs. First, it accurately delineates the location and extent of the lesion. Identification of the pulmonary fissures is helpful to plan the surgical extent. Secondly, and most important in surgical candidates, CT angiography is able to identify systemic arterial supply if present. Appearance reflects the underlying type, and a type III lesion can appear as a consolidation.³ The radiologic differential diagnosis for a solid intrathoracic mass in the neonatal period includes type 3 or fluid-filled CPAM, sequestration, solid (high-grade) pleuropulmonary blastoma, bronchial atresia, and neuroblastoma.⁵

References

1. Gross GW. Pediatric chest imaging. *Curr Opin Radiol* 1992;4(5):36-43
2. Biyyam DR, Chapman T, Ferguson MR, Deutsch G, Dighe MK. Congenital lung abnormalities: embryologic features, prenatal diagnosis, and postnatal radiologic-pathologic correlation. *Radiographics* 2010;30(6):1721-38.
3. Rosado-de-christenson ML, Stocker JT. Congenital cystic adenomatoid malformation. *Radiographics* 1991;11(5):865-86.
4. Berrocal T, Madrid C, Novo S, Gutierrez J, Arjonilla A, Gomez-Leon N. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. *Radiographics* 24(1):e17.
5. Laberge JM, Puligandla P, Flageole H. Asymptomatic congenital lung malformations. *Semin Pediatr Surg* 2005;14(1):16-33.