MDCT spectrum of congenital thoracic anomalies

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Learning objectives

- To learn CT imaging features of the common congenital thoracic anomalies.
- To discuss the CT imaging based differential diagnosis for congenital thoracic anomalies.
Background

Congenital thoracic anomalies are rare, and they may involve the lung parenchyma, diaphragm trachea, oesophagus and mediastinal vasculature. The common congenital chest anomalies include lung agenesis, congenital cystic adenomatoid malformation, congenital diaphragmatic hernia, neuroenteric cyst, and the oesophageal duplication cyst. With advent of MDCT, especially higher detector CT scanners, MPR images obtained help the clinicians to visualise the abnormality better, thus aiding them to plan the surgery and avoid on table surprises.
Findings and procedure details

CT scan of paediatric patients was done with 64 slice multidetector CT scanner (Philips Brilliance, Philips Medical Systems) and were interpreted and accordingly classified into various diseases

1) Lung aplasia:

In pulmonary aplasia is rare congenital anomaly characterized by complete absence of lung parenchyma with rudimentary or hypoplastic bronchus which differentiate it from lung agenesis in which there is complete absence bronchus. It is usually associated other congenital systemic anomaly mainly cardiovascular and in patient of VACTERL association. Contralateral lung and heart occupy the whole of the thorax. There is mediastinal shift to ipsilateral side with displaced aortic arch may compress trachea. Bilateral lung aplasia is incompatible with life.

2) Congenital cystic adenomatoid malformation of lung:

It is developmental hamartomatous abnormality associated with cyst production. It accounts for nearly one fourth of all congenital lung abnormalities. It communicates with the bronchial tree and shows lobar distribution. In early stage it usually presents as a homogeneous, fluid-density mass which later progressing to an air-filled cyst containing multiple air-fluid levels. Initially it may present as one large cyst with mass effect in the form of mediastinal shift to contralateral side. Rapid enlargement of cyst may also occur due to air trapping. On CT scan it commonly presents as small cysts which are usually less than 2 cm in diameter, and may be associated consolidation or larger cysts.

3) Pulmonary sequestration:

Pulmonary sequestration refers to aberrant formation of segmental lung tissue that has no connection with the bronchial tree or pulmonary arteries. Pulmonary sequestration is of two types intralobar and extralobar, depending on its relationship of to the pleura. Intralobar sequestration is more common and usually present later in childhood with recurrent infections. Extra lobar sequestration is less common usually present in early stages with respiratory distress, cyanosis and/or infection. CT scan shows the arterial supply by the descending aorta. It usually doesn’t contain air unless infected. Anomalous arterial supply and venous drainage can be better demonstrated on 3D reconstructions.

4) Congenital Diaphragmatic hernia:
It occurs in 1 of every 2000-3000 live births and accounts for up to 8% of all major congenital thoracic abnormalities. It is of 3 main types: posterolateral: Bochdalek hernia, anterior: Morgagni hernia, hiatus hernia. Left-sided Bochdalek hernias are most common. Left-sided hernias usually have herniation of both small as well as large bowel loops and solid organs such as spleen and left kidney, due to lack of protective effect of liver. On CT scan there is herniation of bowel loops into left thoracic cavity through diaphragm with displacement of lung and mediastinum towards contralateral side.

5) Hypoplastic Right pulmonary artery:

Hypoplasia of the right pulmonary artery is extremely rare. Pulmonary artery hypoplasia should be suspect if a plain chest X-ray shows asymmetric lung fields, lung hypoplasia, or hyperinflation of the contralateral lung in asymptomatic patients. CT scan shows small calibre right pulmonary artery.

6) Tracheal bronchus:

A tracheal bronchus also known as pig bronchus is an anatomical variant where an accessory bronchus originates directly from trachea in supracarinal region. If entire upper lobe is supplied by this bronchus, then it is known as pig bronchus. CT scan gives excellent anatomic information and orientation of tracheal bronchus. Coronal reformation in lung window setting is particularly useful in indentification of tracheal bronchus.

7) Neuroenteric cyst:

Neuroenteric cyst is a type of foregut duplication cyst, which may be associated with vertebral or central nervous system anomalies. They are mainly of two types, 1. Spinal 2. Intra-cranial. Extra medullary intraspinal cysts are more common and they are usually ventral in location. Thoracic region is most commonly affected. The cyst is hypodense on CT scan with soft tissue attenuation. Vertebral abnormality may be associated with neuroenteric cyst.

8) Esophageal duplication cyst:

They occur due to developmental abnormality of the posterior division of embryonic foregut. They usually present as well-defined, thin wall, round or oval shaped, fluid density cystic lesion seen arising from esophagus with cyst contents do not show enhancement. Fat plane with surrounding tissues is maintained.
Fig. 1: Right Lung Aplasia; Image A: Showing the rudimentary cut of the right main bronchus (arrowhead) with non-visualization of the right lung parenchyma. Image B: Showing mediastinal shift to the right with heart in the right hemithorax with compensatory hyperinflation of the left lung herniating in retrosternal space (arrow).

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Fig. 2: Congenital cystic adenomatoid malformation; Image A and B showing multicystic lesion (arrow) in anterior segment of left lower lobe with no obvious communication with segmental bronchus and causing mild mediastinal shift to right.

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Fig. 3: Intralobar pulmonary sequestration; Image A shows mild enhancing soft tissue density mass lesion in anterior segment of right upper lobe and medial segment of right lower lobe (star) receiving systemic blood supply from aorta (arrowhead).

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Fig. 4: Congenital Right Sided hemidiaphragmatic Hernia; Image A: showing herniation of right lobe of liver (black star) into the right hemithorax through peripheral defect in right hemidiaphragm. Image B: anterior section of chest shows herniation of bowel loops through right hemidiaphragm defect with resultant dilatation of bowel loops (arrow head) due to obstruction.

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**Fig. 5:** Isolated hypoplastic right pulmonary artery; Image A showing small calibre right pulmonary artery (arrow) and mildly dilated left pulmonary artery (arrow head).

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Fig. 6: Tracheal bronchus; Image A showing a small bronchus (black arrow head) arising from right side of trachea in supra hilar region supplying right upper lobe in addition to right upper lobe bronchus (white arrow head).

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**Fig. 7:** Neuroenteric cyst; Image A: Contrast enhanced axial CT scan of chest showing large well defined cystic density lesion in posterior mediastinum and extending into left hemithorax (star) causing mass effect in the form of anterior displacement of descending aorta (arrow head). Image B: coronal bone window of chest showing thoracic hemivertebrae (arrow).

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**Fig. 8:** Oesophageal duplication cyst; Image A (axial) and Image B (coronal) CT scan showing well defined cystic lesion in posterior mediastinum bulging into the oesophageal lumen filled with oral contrast (arrow head) and causing luminal narrowing.

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Conclusion

We reviewed 8 cases of congenital malformation of the lung that were studied with computed tomography. We found that CT was helpful in the management of these cases because it helped in confirming the diagnosis, demonstrated unsuspected findings and better depicted anatomic extent on anomalies, thus allowing better planning for surgery. We believe that CT is the method of choice for the study of congenital thoracic especially lung malformations.
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