Case 12385

Congenital segmental bronchial atresia in a young female patient with confirmatory dual energy CT perfusion imaging
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Section: Chest imaging
Area of Interest: Thorax Abdominal wall Respiratory system Lung
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Conventional radiography
Imaging Technique: CT-Quantitative
Special Focus: Congenital Case Type: Clinical Cases
Authors: Twomey M, Ryan J, Moloney F, Maher M
Patient: 36 years, female

Clinical History:

We present a 36-year-old lady with an insidious onset of shortness of breath and abnormal chest X-ray.

Imaging Findings:

An initial chest X-ray at presentation demonstrated an abnormal opacity in the left upper lobe thought to represent infection (Fig. 1). The opacity persisted on follow-up chest X-ray six weeks later (Fig. 2), prompting further evaluation with CT to characterize the lesion.

Contrast-enhanced CT thorax revealed a focal non-enhancing tubular mucocoele with surrounding hyperlucent lung and hyperinflation involving the apico-posterior segment of the left upper lobe (Fig. 3, 4, 5). The remainder of the lung parenchyma was normal.

A subsequent multiplanar dual energy CT lung perfusion study demonstrated increased volume of the left hemithorax with marked segmental hypoperfusion of the apico-posterior segment of the left upper lobe (Fig. 6).

Discussion:

Congenital bronchial atresia (CBA) is a rare condition that occurs when a proximal segmental bronchus is absent, rarely this may occur at a lobar level [1]. The distal bronchial tree is preserved, which leads to under-ventilation and accumulation of mucus in the affected segment [1, 2].

On a chest radiograph, this classically manifests as a focal tubular opacity with a hyperlucent area, which can potentially compress adjacent tissue [2]. Hyperinflation is caused by the unidirectional flow of air from the surrounding lung parenchyma into the affected segment. This is facilitated by tiny inter-alveoli and bronchiolar-alveoli tracts (pores of Kohn and canals of Lambert, respectively) [2, 3]. The absence of a patent bronchus means there is no outflow tract for air or mucus produced by the respiratory epithelium, which leads to the formation of the characteristic bronchocoele/mucocoele. The apicoposterior segment of the left upper lobe is involved most frequently [1]. Of note, CBA and congenital cystic adenomatoid malformation frequently coexist [3].

CT is the most sensitive radiological method for diagnosing CBA. Typical findings include the triad of a mucocoele,
surrounding low attenuation hyperinflated lung and hypoperfusion of the affected segment [1, 2].

Dual energy CT is a relatively novel functional imaging technique, which has predominantly been used in the investigation of pulmonary embolism but is also useful in characterising other causes of lobar or segmental hypoperfusion, as in this case [4].

CBA is generally a benign condition and usually does not require any treatment [2]. It is often discovered incidentally, is typically asymptomatic, and is more common in men. The mean age of diagnosis is 17 years of age [1]. The differential diagnosis includes other abnormalities with mucus impaction, such as allergic bronchopulmonary aspergillosis, cystic fibrosis, or any lesion that causes bronchial narrowing and thus mucus impaction.

Recurrent pulmonary infection is the most common clinical manifestation in symptomatic patients. Bronchoscopy is often required to rule out secondary causes of proximal bronchial obstruction i.e. tumours, foreign bodies or inflammatory strictures [2]. Surgery may be indicated where malignancy cannot be excluded or in the setting of recurrent and severe infection [4]. CBA is suitable for full endoscopic pulmonary resection if surgical intervention is indicated, given the young and otherwise fit demographic group it affects [5].

**Differential Diagnosis List:** Left upper lobe segmental bronchial atresia, Other causes of mucoid impaction: Allergic bronchopulmonary aspergillosis, Endobronchial neoplasm, Arteriovenous malformation, Intralobar sequestration, Congenital lobar emphysema, Intrapulmonary bronchogenic cyst

**Final Diagnosis:** Left upper lobe segmental bronchial atresia

**References:**


Description: Admission chest X-ray demonstrating a tubular opacity in the left upper lobe. A hyperlucent area is seen surrounding the tubular opacity. Origin: Twomey M, Department of Radiology, Cork University Hospital
Description: Contrast-enhanced CT demonstrates focal non-enhancing tubular low attenuation mucocoele in the left upper lobe. Origin: Twomey M, Department of Radiology, Cork University Hospital, Cork, Ireland.
Description: Further axial dual energy CT lung perfusion study demonstrates marked segmental hypoperfusion of the apico-posterior segment of the left upper lobe. Normal perfusion demonstrated in the right upper lobe. Origin: Twomey M, Department of Radiology, Cork University Hospital, Cork, Ireland.
**Figure 4**

**Description:** Follow up chest X-ray 6 weeks later demonstrating persistent left upper lobe tubular opacity (arrow) with surrounding hyperlucent area. **Origin:** Twomey M, Department of Radiology, Cork University Hospital, Cork, Ireland.
Description: Axial contrast-enhanced CT thorax on lung windows shows focal mucocoele (arrow) and surrounding hyperlucent lung in the left upper lobe. Origin: Twomey M, department of Radiology, Cork University Hospital, Cork, Ireland.
Description: Hyperlucent lung noted surrounding the mucocoele (arrow) affecting the apico-posterior segment of the left upper lobe. The remainder of the lung parenchyma is normal. Origin: Twomey M, Department of Radiology, Cork University Hospital, Cork, Ireland.