Clinical challenges

CROSS CANADA ROUNDS FEBRUARY 18, 2016

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Case 1:

- Premature infant with recurrent pneumothorax, despite re-expansion with chest tube placement.
- Chest CT confirms pneumothorax, with multiple cyst-like spaces within parenchyma of one lung

What is the differential diagnosis for cystic lesions in the lung?

Differential diagnosis

Congenital lung diseases

- Single lesions
  - Pulmonary sequestration
  - Bronchogenic cyst
- Multiple lesions
  - Congenital lobar emphysema
  - CPAM
  - Congenital diaphragmatic hernia
  - Lymphangiectasia

Acquired Lung diseases

- Pulmonary interstitial emphysema
- Aspiration
- Infection
- Bronchopulmonary dysplasia
- Langerhans cell histiocytosis

Pulmonary Interstitial Emphysema

- A form of air leak syndrome
- Air escapes from the small airways or alveoli and dissects along the bronchovascular bundles
- Radiate to the periphery to form blebs
  - Blebs burst through visceral pleura → pneumothorax
  - Air passing through the lung hilus → pneumomediastinum
  - Other complications of interstitial air → pneumopericardium, subcutaneous emphysema

Donnelly et al. AJR. 2003; 180

Pulmonary Interstitial Emphysema

- Risk factors include prematurity, surfactant deficiency and low birth weight, meconium aspiration, positive pressure ventilation
- Preterm infants at increased risk
  - Perivascular connective tissue more abundant and less disectable
  - Allows for air trapping in perivascular space
- Different forms:
  - Acute or persistent
  - Localized or diffuse

Bawa et al. Case reports in Pediatrics. 2014; 2014

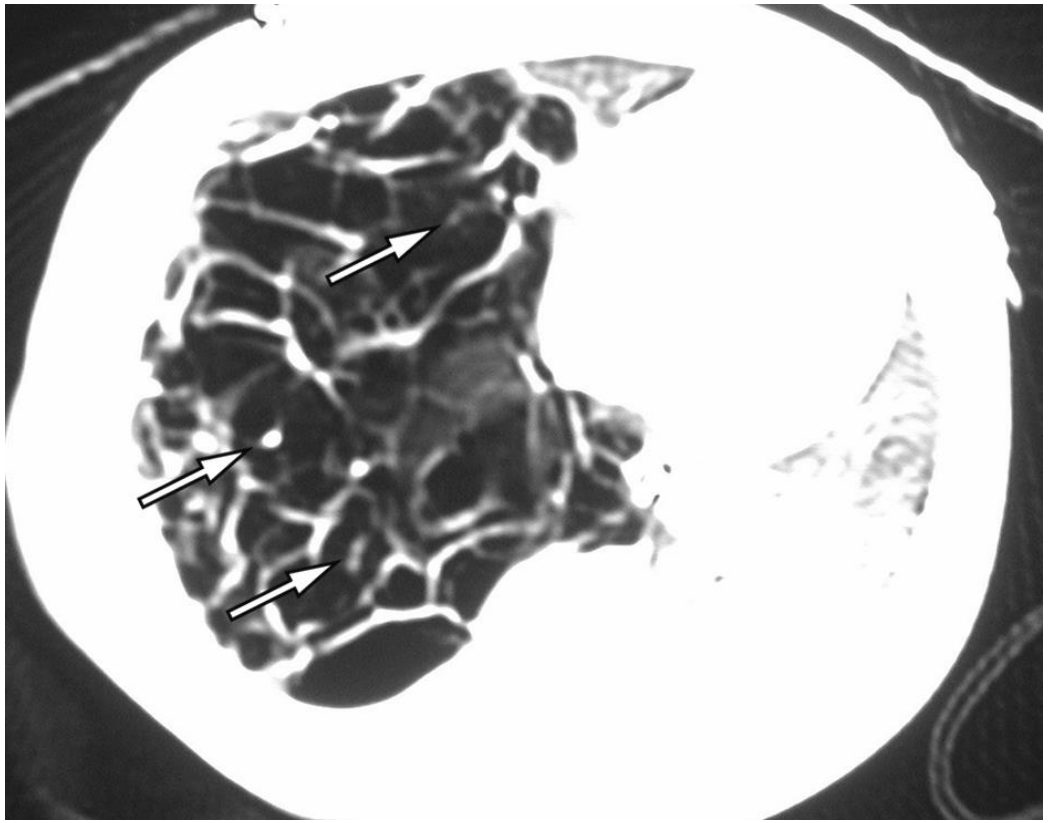
Unusual presentations

- Several cases of PIE in unusual circumstance
  - Never ventilated (Freysdottir et al)
  - CPAP alone (Gurakan et al, Bas et al, Berk et al)
  - Term pregnancy (Bawa et al, Freysdottir et al)
  - Late preterm with higher birth weights (Bas et al)
- These patients all developed localized disease

Freysdottir et al. Pediatric pulmonology. 2006; 41(4)
Gurakan et al. Pediatric Pulmonology. 2002; 34(5)
Berk et al. Pediatric radiology. 2005; 35
Bawa et al. Case reports in Pediatrics. 2014; 2014
Bas et al. Indian pediatrics. 2008; 45(9)

CT scan in PIE

- Multiple cysts
- “Line and dot” pattern
  - Gas in the pulmonary interstitial space surrounding the bronchovascular bundles
  - Present in 82% of persistent localized PIE in one series



Donnelly et al. AJR. 2003; 180

Management of localized PIE

- Lateral decubitus positioning with the affected side down (50-90%)
- Selective bronchial intubation or occlusion
- For more severe disease
  - High frequency ventilation
  - ECMO
  - Lobectomy or pneumonectomy

Donnelly et al. AJR. 2003; 180

Take home points

- Pulmonary interstitial emphysema should be considered in the differential diagnosis of localized cystic lung disease
- It can occur with a history of minimal ventilation, or without a history of ventilation
- CT scan may help to differentiate it from other causes of cystic lung disease
- Conservative measures in localized PIE such as positioning have been found to be curative, and a prolonged trial is recommended provided the patient is stable

Case 2: AO

- 4 year old, previously healthy female referred for possible complicated pneumonia
- Chest X-ray revealed:
  - Left sided pneumothorax
  - Opacity in left lower lobe, with multiple circular lucencies throughout
  - Shift of heart and distal trachea to right
- Chest CT confirmed diagnosis of congenital diaphragmatic hernia
- Patient underwent surgical correction

Late presentation of CDH

- 5-25% of CDH cases
- Associated with a wider spectrum of clinical presentation than neonatal CDH
- Clinical course depends on nature of displaced viscera and its potential complications
- 2:1 male-to-female ratio
- 30% of late presentations over 1 year at diagnosis
  - 23% over 5 years

Baglaj et al. Pediatric Radiology. 2005; 35

Diagnosis

- Chest x-ray only diagnostic in 50% of the cases
  - 14% had “normal” chest x-rays reported in the past
  - In 4% of children, diagnosis was incidental upon investigation for other pathology
  - 36% required serial x-rays, as diaphragmatic hernia not suspected on initial radiograph → some suggestion that herniation may be intermittent
  - Most commonly reported finding: intrathoracic translucent structure associated with mediastinal shift
- 50% required further imaging to make a definitive diagnosis
  - Most common second test in this series was an upper GI contrast study
- Misdiagnosis based on chest radiograph occurred in 25% of patients
  - Pneumonia, pneumothorax, pleural effusion
  - 12% underwent chest tube placement

Baglaj et al. Pediatric Radiology. 2005; 35

Take home points

- Significant number of CDH occur outside the neonatal period
- Clinical and radiographic presentation is variable
  - Symptoms can be very mild
- Often multiple tests required to confirm diagnosis
  - Due to variability in presentation
  - Lack of consideration of diagnosis on practitioner's part

Case 3

- 5 year old Inuit boy from Northern Quebec treated for TB

- On routine follow up, symptoms had abated
  - Chest x-ray revealed cavitary lesion

Cavitary TB

- Rare in young children
  - 2%-7%
  - May be more common in malnourished, immunocompromised children
  - Cavitary TB is a relatively common presentation in adolescents
- Progresses over lengthy period of time i.e. weeks to months
  - In one series, the interval to progression was felt to be quite variable (3-20 months, with most patients falling in the middle)
- Radiographic appearance:
  - Air fluid levels are felt to be rare in TB
- TB treatment failure on direct observed therapy uncommon
  - >95% treatment success

Cruz et al. Pediatrics in review. 2010; 31 (1)
Marais et al. Int J Tuberc Lung Dis. 2004; 8 (4)

Differential diagnosis for cavity

- Infectious
  - Cavitating tuberculosis
  - Cavitating mycosis (*Aspergillus, coccidioides, Histoplasmosis, cryptococcus, PJP*)
  - **Bacterial abscess** (*Anaerobes, Staphylococcus aureus, Klebsiella, Hib*)
  - Hydatid cyst (parasitic)
- Non-infectious
  - Embolism with infarction
  - Vasculitis
  - Pulmonary sequestration
  - Bullae with air-fluid level
  - Localized empyema with air-fluid level

Classification of lung abscess

Primary Lung Abscess

- Normal underlying lung
- Necrotizing pneumonia
- Immunodeficiency
- Aspiration **
  - Dental infection
  - Altered LOC
  - Swallowing disorders, esophageal dysmotility
  - Frequent vomiting, GERD
  - Intubated patients, patients with tracheostomy

Secondary Lung Abscess

- Bronchial obstruction
  - Tumor, enlarged lymph nodes, foreign body, congenital malformation
- Hematogenous dissemination
  - Abdominal sepsis, infective endocarditis, septic thromboembolulism
- Coexisting lung disease
  - Bronchiectasis, Infected pulmonary infarct, contusion, Bullae

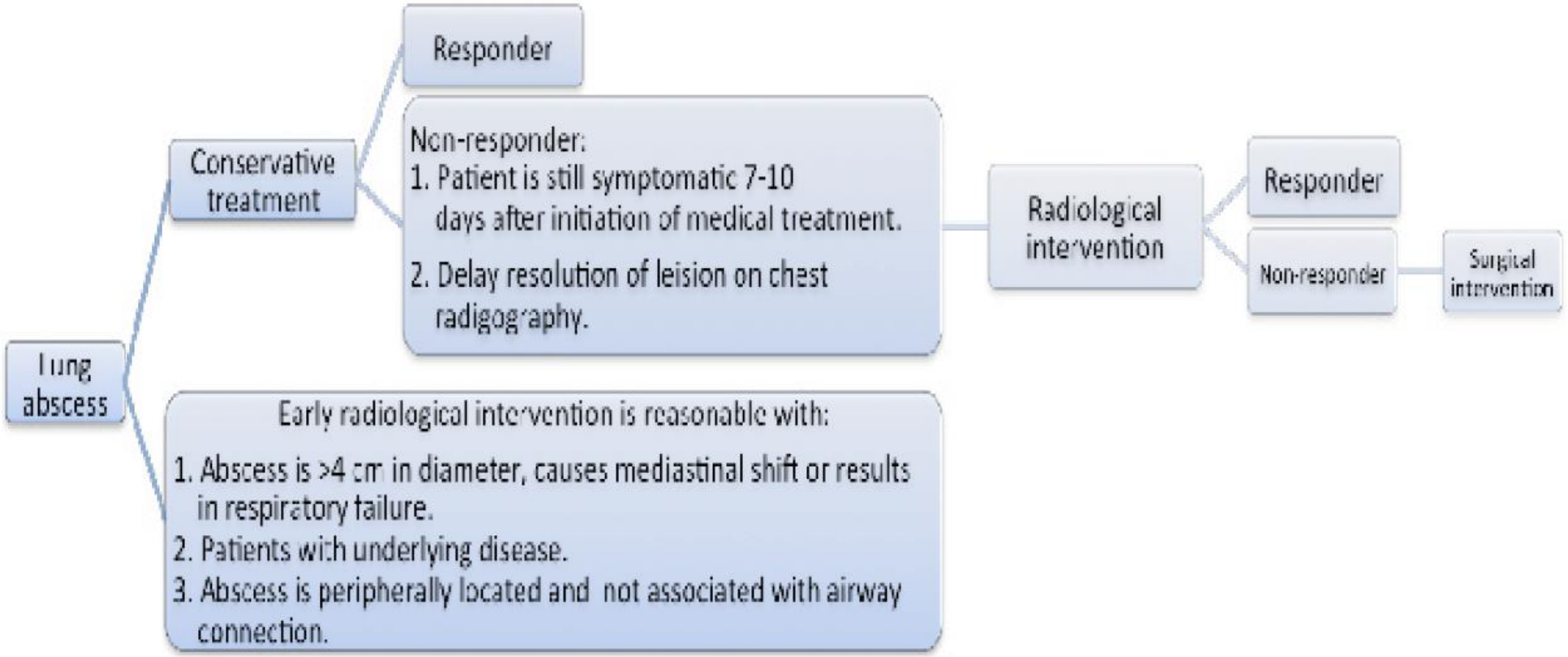
Patradoon-Ho. Paediatric Respiratory Reviews. 2007; 8

Management

- Most pediatric lung abscess will respond to antibiotics
  - Coverage should include *Strep. Pneumoniae, Staph aureus,* Gram negative bacteria
  - Accepted regimens: Cephalosporin + clindamycin, Vancomycin may be considered if MRSA suspected
  - If aspiration suspected, piperacillin-tazobactam, meropenem considered
- No good evidence for duration of treatment
  - Often based on clinical response
  - Parenteral antibiotics often recommended x 2-3 weeks
  - Followed by oral therarpy for 4-8 weeks, until radiographic resolution or stability
- Invasive management may be required if suboptimal response to therapy
  - Interventional radiology, surgery

Lee et al. Journal of Pediatric Respiratory Disease. 2015; 11
Patradoon-Ho. Paediatric Respiratory Reviews. 2007; 8
Yen et al. J Microbiol Immunol Infect. 2004; 37
Kuhajda et al. Annals of Translational Medicine. 2015; 3 (13)

Proposed approach



Lee et al. Journal of Pediatric Respiratory Disease. 2015; 11

Take Home Points

- Cavitary TB is rare in children
- The presence of a lung abscess should prompt consideration of underlying lung disease, immunodeficiency or aspiration
- Most patients will respond to a prolonged course of antibiotics
  - In non-responders, IR intervention or surgery may be necessary

Thank you for your attention

QUESTIONS?