Clinical Summary

- 32 year-old nonsmoker male with life-long history of asthma

- Had 2 hospitalizations during childhood

- Presented to CSMC with progressive dyspnea on exertion for several months, back pain and a non-productive cough
Pulmonary function tests:
- obstructive pattern with significant air trapping
- evidence of small airway disease

V/Q scan severely decreased tracer activity in the right lung which received only 10% of the total lung perfusion and the remaining 90% was from the left lung

CXR showed hyperlucency of the hyperinflated right lung
- Non-functional right lung compressing the opposite lung and the heart
- To further characterized this disease process
- The patient doesn't need it
- Right pneumonectomy
Right middle lobe showing the presence of severe bullous and panacinar emphysema, bronchiectases and peribrochiolar fibrosis.
Right lower lobe showing diffuse changes of panacinar emphysema
Panacinar Emphysema
Peribronchiolar fibrosis
What is the diagnosis?

- Recurrent childhood asthma
- Right lung hyperlucency on CXR
- Non functioning right lung
- Massive unilateral emphysema
- Bronchiectasis
- Peribronchiolar fibrosis
- Pleural bullae
Emphysema

- Panacinar (panlobular) emphysema: acini are uniformly enlarged from the level of the respiratory bronchiole to the terminal blind alveoli.

- Centriacinar emphysema: central and proximal part of the acini are affected; distal alveoli are spared.

- Paraseptal (distal acinar) emphysema: proximal portion of the acinus is normal, but the distal part is predominantly involved.

- Obstructive emphysema: lung expands because air is trapped within it.

- Bullous emphysema: emphysema that produces large subpleural blebs or bullae.
| Differential Diagnosis of Unilateral Lung Hyperlucency |
|---------------------------------|----------------|----------------|----------------|-----------------|----------------|
| Unilateral radiolucency          | Lung volume    | Arterial perfusion | Air trapping | Bronchogram     |
| Sweyer-James-MacLeod syndrome   | Moderate       | Moderate         | Present       | Moderate        | Bronchiectasis |
| Absence of Pulmonary artery     | Slight         | Small            | Absent        | None            | Diminutive bronchial tree |
| Partial obstruction of main bronchus | Marked       | Large            | Present       | Marked          | Lesion in major bronchus |
| Congenital lobar emphysema      | Marked         | Large            | Present       | Marked          | Bronchi displaced around emphysematous area |

Daniel et T et al  Clinical Pediatrics 1984;23:393-397
Swyer-James-MacLeod Syndrome
Historical Background

- First reported case by Swyer and James in 1953 characterized by repeated respiratory infection with right-sided pulmonary hyperlucency on chest X-ray.
- In 1954, MacLeod reported 9 adult cases showing similar features.
- Traditionally known as: Idiopathic unilateral hyperlucent lung or unilateral pulmonary transradiancy.
- “MacLeod’s syndrome” first used by Rivett in 1960.
- “Swyer-James syndrome” first used by Rakower & Moran in 1962.
Swyer-James-MacLeod Syndrome

- Clinical Features:
  - History of recurrent respiratory infections during earlier life
  - Symptomatic patients frequently diagnosed in childhood
  - Asymptomatic patients usually diagnosed later in life when CXR is done for other reasons
    - Increasing dyspnea, tightness of the chest, chronic cough and easy fatigability

- Characteristic Radiographic Findings:
  - Unilateral pulmonary hyperlucency on CXR secondary to a decrease in vascular shadows over the lung field
Etiology

- Etiology and pathogenesis remain unknown

- Yet to be determined whether congenital or acquired

- Two theories raised regarding its pathogenesis:
  A. Bronchial Pathogenic Theory
  B. Pulmonary Artery Pathogenic Theory
Bronchial Pathogenic Theory

- Bronchioles are first obstructed due to pulmonary infection
- Emphysematous change takes place leading to dilated alveoli
- Subsequently press pulmonary vessels
- Decreased pulmonary blood supply
- Congenital dysplasia of pulmonary artery
- Inhibiting the bronchi growth
- Making emphysematous changes inevitable along with bronchiolitis
Other pathologic findings: What is this?
Placental transmogrification

Transmogrification means to change into different form or shape especially one that is fantastic or bizarre:

- First described by McChesney in 1979 in a case report
- Morphologically bears superficial resemblance to placental villi
- Represents a rare disease with only 23 cases reported in the literature to date
- Adults and young adults, predominantly male with history of pneumonic process

- All cases have been described in patients with intrapulmonary cystic lesions, may appear as a bulla or enlarging cyst

- Different opinions postulated on its pathogenesis but no single explanation
Different opinions on its pathogenesis

- Represents primarily a benign proliferation of peculiar interstitial clear cells with secondary cystic changes (Cavazza et al)

- Its presence in patient with emphysema maybe the result of metaplastic mesenchymal differentiation (Hochholzer et al)

- Related to chronic segmental edema due to destruction or blockage of the lymphatics in the affected areas (Koss, M)

- May have been induced by proliferation of the epithelial cells lining the hamartomas (Xu et al)

- Our opinion, probably results from the development of edema, fibrosis and chronic inflammation in the residual alveolar tissue of patients with severe emphysema
Conditions associated with Placental Transmogrification

- Severe cigarette-smoking induced emphysema
- Giant bullous emphysema of the lung
- Fibrochondromatous hamartomas
- Ours is the first case associated with Swyer-James-MacLeod syndrome
Swyer-James-MacLeod Syndrome with Placental Transmogrification
References:

- Swyer PR, James GCW. A case of unilateral pulmonary emphysema. Thorax 1953; 8: 133-6