# Diseases of the Respiratory System

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## Atelectasis

- 1. Definition: area of collapsed or nonexpanded lung
- 2. Major types
  - a. Obstruction/resorption atelectasis
    - i. Collapse of lung due to resorption of air distal to an obstruction
    - ii. Examples: aspiration of a foreign body, chronic obstructive pulmonary disease(COPD), or postoperative
  - b. Compression atelectasis due to fluid, air, blood, or tumor in the pleural space
  - c. Contraction (scar) atelectasis due to fibrosis and scarring of the lung
  - d. Patchy atelectasis
    - i. Due to a lack of surfactant
    - ii. Examples: hyaline membrane disease of newborn or acute (adult) respiratory distress syndrome (ARDS)
- 3. Predisposed to infection
- 4. Reversible disorder

## Bacterial pneumonia

- a. Definition: acute inflammation and consolidation (solidification) of the lung due to a bacterial agent
- b. Clinical signs and symptoms
  - i. Fever and chills
  - ii. Productive cough with yellow-green (pus) or rusty (bloody) sputum
  - iii. Tachypnea
  - iv. Pleuritic chest pain
  - v. Decreased/Increased breath sounds, rales, and dullness to percussion
- c. Lab: elevated WBC *count with a left shift*
- d. Chest x-ray
  - i. Lobar: lobar or segmental consolidation (opacification)
  - ii. Bronchopneumonia: patchy opacification
  - iii. Pleural effusion
- e. Clinical keys: identification of the organism and early treatment with antibiotics

- f. Lobar pneumonia
  - i. Consolidation of entire lobe
  - ii. Organism: *Streptococcus pneumoniae (95%) or Klebsiella*
  - iii. Four classic phases
    - . Congestion: active hyperemia and edema
    - . Red hepatization: neutrophils and hemorrhage
    - . Grey hepatization: degradation of red blood cells
    - . Resolution: healing
  - iv. Micro: intra-alveolar supporative inflammation (neutrophils) and edema
- g. Bronchopneumonia
  - i. Scattered patchy consolidation centered around bronchioles
  - ii. Tends to be bilateral, multilobar, and basilar
  - iii. Affects the young, old, and terminally ill
  - iv. Organism: Staphylococci, Streptococci, *Haemophilus influenzae, Pseudomonas aeruginosa,etc.*
  - v. Micro: acute inflammation of bronchioles and surrounding alveoli

- h. Diagnosis
  - i. Sputum gram stain and culture
  - ii. Blood cultures
- i. Treatment: empiric antibiotic treatment modified by the *results of cultures and* organism sensitivities
- j. Complications of pneumonia
  - i. Fibrous scarring and pleural adhesions
  - ii. Lung abscess(pus)
  - iii. Empyema(accumulation of pus)
  - iv. Sepsis(presence of microorganism or toxins in tissue or bloodstream)

## Lung abscess

- a. Definition: localized collection of neutrophils (pus) and necrotic pulmonary parenchyma
- b. Etiology
  - i. Aspiration .
    - Most common
    - Tends to involve right lower lobe
    - Mixed oral flora (anaerobic/aerobic)
  - ii. Following a pneumonia, especially S. *aureus and Klebsiella*
  - iii. Postobstructive
  - iv. Septic emboli
- c. Complications
  - i. Empyema
  - ii. Pulmonary hemorrhage
  - iii. Amyloidosis

## Sarcoidosis

- 1. Epidemiology
  - a. Unknown etiology
  - b. Females> males, age: 20-60
  - c. Most common in African American women
- 2. Clinical presentation
  - a. May be asymptomatic
  - b. Cough, shortness of breath (SOB)
  - c. Fatigue, malaise
  - d. Skin lesions
  - e. Eyeirritation or pain
  - f. Fever/night sweats

- 3. Noncaseating granulomas occur in any organ of the body
  - a. Lung: diffuse scattered granulomas
  - b. Lymph nodes: hilar and mediastinal adenopathy
  - c. Skin, liver/spleen, heart, CNS
  - d. Eye:Mikulicz syndrome: involvement of uvea and parotid
  - e. Bone marrow: especially in the phalanges
- 4. Lab: elevated serum *angiotensin convertingenzyme (ACE)*
- 5. X-ray: bilateral hilar lymphadenopathy
- 6. Micro
  - a. Noncaseating granulomas
  - b. *Schaumann bodies:laminated calcifications*
  - c. *Asteroid bodies: stellate giant-cell cytoplasmic inclusions*
- 7. Diagnosis of exclusion
- 8. Prognosis: favorable with a variable clinical course

## Obstructive Versus Restrictive Lung Disease

| Obstructive | Restrictive |
|---|---|
| Definition:<br>Increased resistance to airflow secondary to obstruction of airways | Decreased lung volume and capacity |
| Pulmonary function tests (spirometry)<br>FEVl/FVC ratio is decreased | Decreased TLC and VC |
| Eg:<br>Asthma<br>Chronic Bronchitis<br>Emphysema<br>Bronchiectasis | <u>Chest wall disorders</u><br>Obesity, kyphoscoliosis, polio, etc<br><br><u>Interstitial/infiltrative diseases</u><br>ARDS,pneumoconiosis . Pulmonary fibrosis |

## Chronic Obstructive Pulmonary Disease (COPD)

- 1. Chronic bronchitis
  - a. Clinical diagnosis: persistent cough and copious sputum production for at least 3 months in 2 consecutive years
  - b. Highly associated with smoking *(90%)*
  - c. Clinical findings
    - i. Cough, sputum production, dyspnea, frequent infections
    - ii. Hypoxia, cyanosis,weight gain
  - d. Micro
    - i. Hypertrophy of bronchial mucous glands (Reid index)
    - ii. Increased numbers of goblet cells
    - iii. Hypersecretion of mucus
    - iv. Bronchial squamous metaplasia and dysplasia (smokers)

  - e. Complications
    - i. Increased risk for recurrent infections
    - ii. Pulmonary HTN leading to right heart failure (cor pulmonale)
    - iii. Lung cancer
- 2. Emphysema
  - a. Definition: destruction of alveolar septa resulting in enlarged air spaces and a loss of elastic recoil
  - b. Etiology
    - i. Protease/antiprotease imbalance
    - ii. Proteases (including elastase) are produced by neutrophils and macrophages, which are stimulated by smoke and pollution
    - iii. Antiproteases include a-I-antitrypsin, a-I-macroglobulin, and secretory leukoprotease inhibitor
    - iv. Centriacinar Vs Panacinar

  - c. Gross
    - i. Overinflated, enlarged lungs
    - ii. Enlarged, grossly visible air spaces
    - iii. Formation of apical blebs and bullae (centriacinar type)
  - d. Clinical findings
    - i. Progressive dyspnea
    - ii. Pursing of lips and use of accessory muscles to breathe
    - iii. Barrel chest
    - iv. Weight loss

## Asthma

- a. Definition: hyperreactive airways, resulting in episodic bronchospasm when triggered by certain stimuli
- b. Extrinsic (type I hypersensitivity reaction)
  - i. Allergic (atopic)
    - . Most common type
    - . Childhood and young adults; (+) family history
    - . Allergens: pollen, dust, food, molds, animal dander, etc.
  - ii. Occupational exposure: fumes, gases, and chemicals

- c. Intrinsic (unknown mechanism)
  - i. Respiratory infections (usually viral)
  - ii. Stress
  - iii. Exercise
  - iv. Cold temperatures
  - v. Drug induced (aspirin)
- d. Asthma attack: wheezing, severe dyspnea, coughing
- e. Status asthmaticus: potentially fatal unrelenting attack
- f. Sputum cytology
  - i. *Curschmann spirals:twisted mucous plugs admixed with sloughed epithelium*
  - ii. Eosinophils
  - iii. *Charcot-Leydencrystals:eosinophil membrane protein*

- g. Micro
  - i. *Mucous plugs*
  - ii. Hypertrophy of mucous glands with goblet cell hyperplasia
  - iii. Inflammation (especially with eosinophils)
  - iv. Edema
  - v. Hypertrophy of bronchial wall *smooth muscle*
  - vi. Thickened basement membranes

## Bronchiectasis

- a. Definition: abnormal permanent airway dilatation due to chronic necrotizing infection
- b. Cough, fever,malodorous purulent sputum, dyspnea
- c. Causes
  - i. Bronchial obstruction: foreign body, mucous, *tumor, etc.*
  - ii. Necrotizing pnuemonias
  - iii. Cystic fibrosis

  - iv. Kartagener syndrome .
    - Autosomal recessive
    - . Immotile cilia due to defect of dynein arms
    - . Bronchiectasis, sinusitis, situs inversus
- d. Gross: dilated bronchi and bronchioles extending out to the pleura
- e. Complications: abscess, septic emboli, cor pulmonale, amyloidosis

## Adult respiratory distress syndrome (ARDS)

- a. Synonyms: diffuse alveolar damage (DAD), shock lung
- b. Definition: diffuse damage of alveolar epithelium and capillaries, resulting in progressive respiratory failure that is unresponsive to oxygen treatment
- c. Causes: shock, sepsis, trauma, gastric aspiration, radiation, oxygen toxicity, drugs, pulmonary infections, and many others
- d. Clinical presentation: dyspnea, tachypnea, hypoxemia, cyanosis, and use of accessory respiratory muscles
- e. X-ray: bilateral lung opacity ("white out")
- f. Gross: heavy,stiff, noncompliant lungs
- g. Micro
  - i. Interstitial and intra-alveolar edema
  - ii. Interstitial inflammation
  - iii. Loss of type I pneumocytes
  - iv. Hyaline membrane formation
- h. Treatment
  - i. Treat the underlying cause
  - ii. Oxygen, positive end-expiratory pressure (PEEP), and mechanical ventilation
  - i. Prognosis: overall mortality 50%

## Respiratory distress syndrome of the newborn

- a. Synonym: hyaline membrane disease of newborns
- b. Associated with
  - i. Prematurity (gestational age of <28 weeks has a 60% incidence)
  - ii. Maternal diabetes
  - iii. Multiple births
  - iv. C-section delivery
- c. Defect: *deficiencyof surfactant*
- d. Clinical presentation: often normal at birth, but within a few hours develop increasing respiratory effort, tachypnea, nasal flaring, use of accessory muscle of respiration, an expiratory grunt, cyanosis
- e. X-ray: "ground-glass" reticulogranular densities
- f. Lab: lecithin:sphingomyelin ratio <2
- g. Micro: atelectasis and *hyaline membraneformation*
- h. Treatment: surfactant replacement and oxygen
- i. Prognosis: overall mortality ~30%

- J. Complications of oxygen treatment in newborns
  - i. Bronchopulmonary dysplasia
  - ii. Retrolental fibroplasia (retinopathy of prematurity)
- k. Prevention: delay labor and corticosteroids to mature the lung

## Pulmonary edema

- a. Definition: fluid accumulation within the lungs usually due to disruption of Starling forces or endothelial injury
- b. Increased hydrostatic pressure: left-sided heart failure, mitral valve stenosis, fluid overload
- c. Decreased oncotic pressure: nephrotic syndrome, or liver disease
- d. Increased capillary permeability: infections, drugs (bleomycin, heroin), shock, radiation
- e. Gross: wet, heavy lungs; usually worse in lower lobes
- f. Micro: intra-alveolar fluid, engorged capillaries, hemosiderin-laden macrophages (heart -failure cells)

## Pulmonary emboli (PE) and pulmonary infarction

- a. Most (90-95%) pulmonary emboli arise from deep vein thrombosis (DVT) in the leg
- b. Only 10% of pulmonary emboli cause infarction
- c. Infarcts usually occur in patients with *underlying cardiopulmonarydisease*
- d. Gross: wedge-shaped, hemorrhagic infarction
- e. Diagnosis:V/Q lung scan ~ V/Q mismatch
- f. Complications
  - i. Large emboli (saddle emboli) may cause sudden death
  - ii. Septic emboli may result in a pulmonary abscess

## Pulmonary hypertension

- a. Definition: increased pulmonary artery pressure, usually due to increased vascular resistance or blood flow
- b. Etiology
  - i. COPD and interstitial disease (hypoxic vasoconstriction)
  - ii. Multiple ongoing pulmonary emboli
  - iii. Mitral stenosis and left heart failure
  - IV. Congenital heart diseasewith left to right shunts (ASD,VSD,PDA)
  - v. Primary (idiopathic)
- c. Pathology
  - i. Pulmonary artery athlerosclerosis
  - ii. Small artery medial hypertrophy and intimal fibrosis
  - iii. Plexogenic pulmonary arteriopathy
  - iv. Complication: right ventricular hypertrophy ~ failure (cor pulmonale)