Imaging of Pulmonary Hemorrhage Syndromes

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Objectives

• Recognize patterns of pulmonary hemorrhage
• List the leading causes of pulmonary hemorrhage
• Include pulmonary hemorrhage in the differential diagnosis of other disease processes

CR patterns of pulmonary hemorrhage

• Diffuse pulmonary hemorrhage or DPH
  – Patchy or diffuse opacities
  – Consolidation
  – Perihilar distribution

Histologic basis of pulmonary hemorrhage

• Vasculitis or Capillaritis
  – Neutrophil infiltration of the interstitium leading to anatomic disruption of the capillaries and extravasation
• ‘Bland’ pulmonary hemorrhage
  – Extravasation without inflammation or destruction of vessels
• Diffuse alveolar hemorrhage associated with another condition
  – Diffuse alveolar damage
  – Lymphangioleiomyomatosis

CT patterns of pulmonary hemorrhage

• Diffuse pulmonary hemorrhage
  – Ground glass opacities and consolidation
  – Ill-defined centrilobular nodules
  – Interlobular septal thickening
  – Crazy paving pattern
Histologic patterns of DPH

- Vasculitis or capillaritis
  - Granulomatosis with polyangiitis - GPA (Wegener granulomatosis)
  - Microscopic polyangiitis - MPA
  - Antiglomerular basement membrane disease - AGBM (Goodpasture syndrome)
  - Collagen vascular disease
  - Acute lung graft rejection
  - others...

- 'Bland' pulmonary hemorrhage (w/o vasculitis or capillaritis)
  - Toxins or illicit drug use (crack lung)
  - Idiopathic pulmonary hemosiderosis
  - Pulmonary veno-occlusive disease - PVOD
  - Cardiac valvular disease (MR/MS)

Histologic patterns of DPH

- Hemorrhage associated with another condition
  - Diffuse alveolar damage - DAD
  - Pulmonary embolism
  - Barotrauma
  - Pulmonary capillary hemangiomatosis - PCH

Leading causes of DPH to be discussed

- Granulomatosis with polyangiitis - GPA (WG)
- Antiglomerular basement membrane disease – AGBM (Goodpasture syndrome)
- Connective tissue disease – SLE
- Idiopathic pulmonary hemosiderosis
- Pulmonary venous occlusive disease (PVOD)

Granulomatosis with polyangiitis

- Unknown cause
- Upper and lower respiratory tract
- Glomerulonephritis
- Necrotizing vasculitis

Granulomatosis with polyangiitis

- Neutrophilic capillaritis with hemorrhage
- Vascular occlusion
- Lung inflammation
Granulomatosis with polyangiitis
- Consolidation and ground glass opacities (hemorrhage)
- Pulmonary nodule or masses, sometimes cavitary
- Solitary nodule or mass

Antiglomerular basement membrane disease
- Young patients, 20-30 year olds
- M>F, four times
- Hemoptysis and anemia, ~90%
- Renal disease common

Antiglomerular basement membrane disease
- Serum antibodies in 95%
- Pulmonary capillaritis
- Glomerulonephritis

Antiglomerular basement membrane disease
- Diffuse airspace or groundglass opacities
- Bilateral and symmetric
- Perihilar distribution

Collagen vascular disease
- Most frequent in SLE
  - Uncommon initial presentation
  - Hemoptysis massive and frequent
  - High mortality

Collagen vascular disease
- Consolidation and ground glass opacities
- Crazy paving pattern
- Centrilobular and perivascular opacities
  - Perivascular inflammation and hemorrhage
Idiopathic pulmonary hemosiderosis

- Young patients, aged <10
- Episodic pulmonary hemorrhage
- Association with celiac disease, IgA gammopathy
- Responds to a gluten free diet?

Pulmonary venous occlusive disease - PVOD

- Similar to PCH
- Variant of pulmonary hypertension
- Usually fatal in just a few years

Additional causes of pulmonary hemorrhage

- Microscopic polyangiitis
- Paucy-immune pulmonary capillaritis
- Henoch-Schönlein purpura
- Antiphospholipid syndrome
- Behçet syndrome
- Thrombotic or idiopathic thrombocytopenic purpura
- Mitral stenosis or regurgitation
- Toxins/illicit drug use
- Diffuse alveolar damage
- Barotrauma
- And so on...

Differential diagnosis

- Other similar appearing entities
  - Pulmonary edema
    - Cardiogenic and noncardiogenic
  - Community acquired pneumonia
  - Pneumocystis jiroveci
  - Mycoplasma pneumonia
  - Septic emboli
  - Could be confused with GPA
  - Drug reaction
Summary

- We reviewed several patterns of pulmonary hemorrhage
- We reviewed leading causes of pulmonary hemorrhage
  - Granulomatosis with polyangiitis - GPA (WG)
  - Antineutrophil cytoplasmic antibody disease - ANCA (Wegener’s granulomatosis)
  - Connective tissue disease – SLE
  - Idiopathic pulmonary hemosiderosis
  - Pulmonary venous occlusive disease (PVOD)
- We listed other common diseases with a similar pattern
  - Pulmonary edema
  - Community-acquired pneumonia
  - Pneumocystis jiroveci pneumonia
  - Drug reaction

References