Pathology of pulmonary vascular disease

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Pulmonary vascular disease

Type of pulmonary circulation:

Types of pulmonary vascular disease

Objectives
Pathology of Pulmonary Vascular Diseases
To discuss the etiology, morphological features and clinical consequences of **Pulmonary embolism (PE)**.

To describe the pathogenesis, morphology and clinical features of **Pulmonary hypertension (PH)**.

**Pulmonary embolism (PE)**

**Definition:**
- Impaction of a thrombus or foreign matter in the pulmonary
vascular bed as secondary of other conditions, leads to complications and death.

► **Process:**
► Blood clots formation → **BREAK & TRAVEL** to occlude the pulmonary arteries & branches (one or more).

► **Types:** (1) Thrombotic (2) Non-thrombotic

► **Source of Non-thrombotic PE (rare):**
► 1. Tumors,
► 2. Air bubbles. 3. Amniotic fluid.
► 4. Fat.
The venous thromboembolism (VTE) refers to DVT, PE, or to a combination of both.

Rudolf Virchow "Father of Pathology"

(>90%) of PE cases are originating from the deep veins e.g popliteal vein.
All predisposing factors to DVT is well explained by him.

Virchow-triad
- Stasis of blood flow.
- Endothelium Injury (irritation, trauma)
- Hypercoagulability (Thrombophilia).

PE Predisposing/Risk factors:
► **Inherited** Hypercoagulable states, (AT III def., protein C, S deficiency).

► **Acquired**

► **Immobilization**- Bed rest

► **Post-operative** (Hip, legs, abdomen)

► Severe blood loss and trauma (fractures & burns)

► **Women** (Pregnant, oral contraceptive rich in ER)

► Varicose veins

► Advancing age.

► Obesity, smoking
Malignancy

DM

Cardiac diseases- CHF, HTN, MI, Fibrillation

Primary polycythemia.

Race

PE morphology-

Origin

1. Thrombotic in origin- most common.

2. Veins > Arteries.
3. Typical sites: **Deep veins of the calf and Deep pelvic veins**

- **Large-vessel in situ thromboses** are rare.
- **PE morphology**
  - may lodge in various sites in the pulmonary arterial tree.
  - **1) Large emboli** lodge the in the main pulmonary artery or its major branches or at the bifurcation as a **saddle embolus** (sudden death).
PE morphology based on site and size

2) Hemorrhages at the periphery (small emboli).

3) Lung infarction- Wedge shaped, (base at the pleural surface & the apex pointing to the hilus of the lung)- hemorrhagic.

4) Thrombus/clot can be distinguished from a post-mortem clot by the presence
of the lines of Zahn in the thrombus.

- **Microscopic of pulmonary infarct**
  - Ischemic necrosis of the lung within the area of hemorrhages, alveolar, bronchioles, BV
• **Cellular events with Hemosiderin deposits**

3) Infected embolus, reveals intense neutrophilic inflammatory reaction referred as septic infarcts= abscesses.

4) Fibrous replacement – converts into a contracted scar.
PE morphology-based on emboli source

PE- Clinical course

- 60-80% are clinically silent.
- 5% sudden death (large emboli).
- < 3% of cases recurrent pulmonary infarcts, result in pulmonary HTN & RIGHT HF.
Common symptoms & signs

Diagnosis: D-DIMER, USG-DOPPLER, CT, MRI

PE - The clinical effect & Consequences
The clinical effect depends on
Two main pathophysiologic “effects” consequences:

- PE - outcome
1) Occlusion of a major vessels leads to:

   I. Sudden DEATH →
   (Unresolved + complication, HF)

2) Occlusion of a smaller vessel:

   I- No effect if the bronchial circulation is good. (resolved)
   II - Pulmonary HT → (If small and multiple +Recurrence).
   III. Pulmonary infarction
► PULMONARY HYPERTENSION (PH) objectives

► To describe the pathogenesis, morphology and clinical features of pulmonary hypertension (PH).

► WHAT IS PULMONARY HYPERTENSION

► Definition:
is Hemodynamic **SERIOUS & FATAL** illness chr. by **high BP** in the affected **BV** in the lungs and **Right side** of the heart - due to **narrowed, blocked or destroyed** **BV**.

**BV** = Pulmonary Arteries, capillaries & veins.

The mean pulmonary artery pressure (mPAP) reaches BP > 25 mm Hg at rest & > 30 mm Hg during exercise. (measured by right heart catheterization).

**PH** - isn't curable, treatments are available that can help lessen symptoms and improve quality of life.

**Complications** →

RHF, CLOT, BLEEDING (hemoptysis), Arrhythmia
What’s the main causes of PH?

The pressure in the lung BV increased for two reasons:

1) Increased blood flow.
2) Increased resistance within the pulmonary circulation. (narrowed, destroyed, blocked)

Can be classified into three main causes based on etiology:

- **Secondary pulmonary hypertension**:
- caused by another medical problem, e.g.
• PE, CT disease, Sickle cell anemia
• COPD, Lung fibrosis & scarring, HIV, Drugs-induced.
• Cardiac diseases, LHF, vasculitis.

• 2- **Primary pulmonary hypertension (Familial)**
  - Rare, **Mutations, autosomal dominant inheritance.**
    - No underlying cause. Patients are rather sensitive to any vasoconstrictors.

► **3) Idiopathic PH:**
► Sporadic, requires exclusion of others.
► Usually women 20-40 years old, some time children.
PH- Pathogenesis

- Occurs in Primary PH (familial) →
  Mutations in the bone morphogenetic protein receptor type 2 (BMPR2) → BV thickening & occulsion.

- Occurs in Secondary PH → produced endothelial cell dysfunction e.g.- Leftto-right shunts (Mechanical), Thrombo-embolism, (biochemical injury produced by fibrin).

- Occurs in Secondary PH → Platelet Aggregation & adhesion + Endothelial activation + Cytokines production + vasospastic effect.
PH morphology

1. Medial hypertrophy

2. Atheromatous deposits.

3. Intimal fibrosis-narrowing

4. Organizing or recanalized thrombi, with coexistence of diffuse fibrosis this favors recurrence.

5. Alveolar hemorrhages
Morphology of PH-Gross changes

Pulmonary hypertension, reveal atheroma formation, usually limited to large vessels

6- Plexiform lesion-in small arteries multichannel.

- Associated with:
  - Idiopathic & primary PH+
Congenital heart disease with left-to-right shunts.

PH Clinical features

Signs & symptoms:

Like HTN are subtle in the early stages.

Hidden by underlying diseases.

Varying from pt. to pt.

Initial Symptoms: dyspnea, cough, fatigue, chest angina-like pain, slowed growth (in child).

Overtime Severe respiratory distress, cyanosis, and right ventricular hypertrophy, RHF.
PH outcome: Death from decompensated cor pulmonale, often with superimposed thromboembolism and pneumonia.

THE END