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Pathology of pulmonary vascular disease

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- **Pulmonary vascular disease**
- **Type of pulmonary circulation:**

- **Types of pulmonary vascular disease**

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Objectives

<u>Pathology of Pulmonary Vascular Diseases</u>

- To discuss the etiology**1**, morphological**2** features and clinical consequences of P**ulmonary embolism(PE)**

- To describe the pathogenesis**1,**morphology**2** and clinical features**3** of P**ulmonary 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)
  - **Hypercoagulablity** (Thrombophilia).

- **<u>PE Predisposing\Risk factors:</u>**

- **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

- 1ry 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 <u>narrowed</u>, <u>blocked</u> or <u>destroyed</u> 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.
- **<u>Complications</u>→**
  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- <u>Primary pulmonary hypertension (Familial)</u>**

- Rare, **Mutations, autosomal dominant inheritance**.
- No underlying cause. Patients are rather sensitive to any vasoconstrictors.

- **3) <u>Idiopathic PH:</u>**
- Sporadic, requires exclusion of others.
- Usually women 20-40 years old, some time children.

▶PH- Pathogenesis

- **Occurs in Primary PH(familial)→** **M**utations 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.Initimal 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**
- **<u>Sign& symptoms:</u>**
- Like HTN are **subtle** in the early stages**.**
- **Hidden** by underlying diseases.
- **Varying** from pt. to pt.

- **<u>Initial Symptoms</u>:** dyspnea, cough, fatigue, chest angina-like pain, slowed growth (in child)**.**
- **<u>Overtime</u> Severe respiratory distress**, **cyanosis**, and **right ventricular hypertrophy, RHF.**

- **PH outcome:** **Death** from decompensated cor pulmonale, often with superimposed thromboembolism and pneumonia.

THE END