



Why is ARDS in all CAPS?

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Introduction

The differential for acute bilateral pulmonary infiltrates includes cardiogenic edema, infection, inflammation, and alveolar hemorrhage. Acute respiratory distress syndrome (ARDS) diagnosis is based on the **Berlin criteria**:

1. Hypoxemia; defined as arterial oxygen (PaO₂) to fraction of inspired oxygen (FiO₂) of <300mmHg on mechanical ventilation with PEEP >= 5 mmHg
2. Onset within one week
3. Bilateral infiltrates on chest xray or CT
4. Symptoms not explained by heart failure

Case

Patient: Middle-aged male

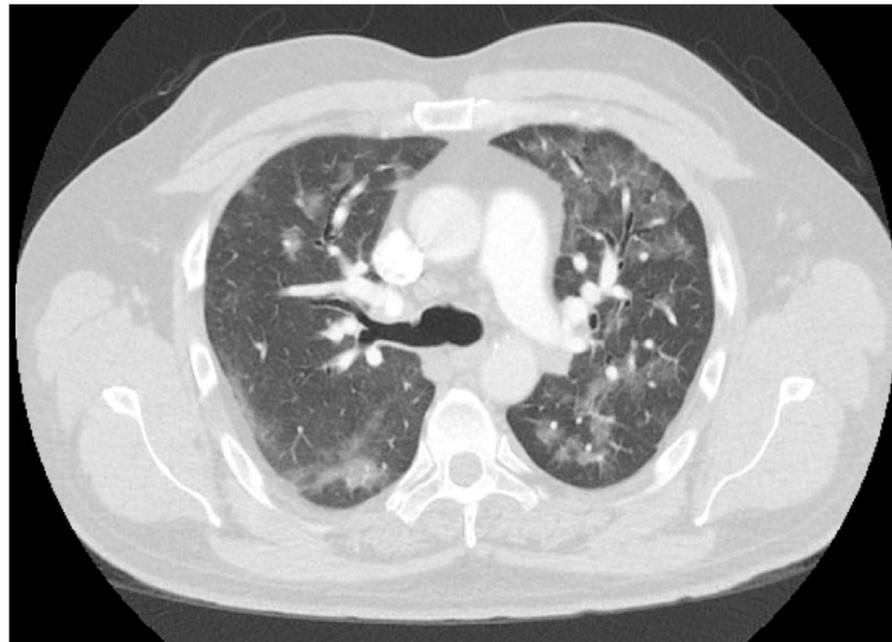
PMH: Peripheral artery disease s/p right below knee amputation on enoxaparin and antiphospholipid syndrome (APS)

CC: Red painful rash of his right leg with increased pain

Course: Dyspneic at transferring hospital, placed on 2L NC. CT-PE negative for pulmonary embolism but showed multifocal ground glass opacities. He was given 2 liters of crystalloid and started on piperacillin/tazobactam and azithromycin. Ultrasound of right leg was negative for blood clots. Transfer was initiated to tertiary care hospital where he follows with Vascular Surgery.

Case (cont.)

Upon presentation to tertiary hospital, vitals 38.5 °C, HR 125, BP 106/77, Resp 24 and SpO₂ 86 % on 6LNC. Placed on heated high flow nasal cannula 40LPM/100% for **persistent hypoxemia**. Chest xray showed bilateral diffuse infiltrates and CBC showed leukocytosis as well as new anemia. Other labs notable for Troponin T of 1212 and elevated inflammatory markers. CT femur showed possible cellulitis but negative for abscess. He was intubated for hypoxemic respiratory failure with ABG 7.36/44/125 on 80% FiO₂ (**P/F=156**) and transferred to the ICU with multiple specialists on consultation.



Conclusions

- Recognize ARDS and implement **lung protective ventilation** (tidal volume of ≤6 ml/kg ideal body weight) if needed.
- There is a **wide differential** for pulmonary infiltrates on chest xray including cardiogenic edema, infection, inflammation, and alveolar hemorrhage.
- CAPS is an uncommon but life threatening cause of respiratory failure and is managed with supportive care, anticoagulation, steroids, plasma exchange, IVIG, and possibly chemotherapy.

Discussion

The patient presented with acute hypoxic respiratory failure concerning for ARDS. He was diagnosed with **catastrophic antiphospholipid antibody syndrome (CAPS)** based on multi organ system involvement and biopsy. Differential on ED presentation was wide.

<i>DDx for bilateral pulmonary infiltrates</i>	
<i>Cardiogenic pulmonary edema</i>	<i>Bilateral pneumonia</i>
<i>Diffuse alveolar hemorrhage</i>	<i>Vasculitis</i>
<i>Eosinophilic pneumonia</i>	<i>CAPS</i>
<i>Malignancy</i>	<i>Cryptogenic org pneumonia</i>

Mortality is CAPS approaches 30% and early treatment is critical. It is characterized by widespread thrombotic disease and organ failure. This patient had biopsy of his leg rash was consistent with superficial thrombosis. With concomitant respiratory failure and renal failure, he met criteria for CAPS. There is **often an inciting event**, such as amputation site infection in this patient.

Management of CAPS:

1. **Antibiotics if inciting infection**
2. **Respiratory support**
3. **High dose steroids**
4. **Anticoagulation**
5. **Plasma exchange ± IVIG**
6. **± Chemotherapy**