Systemic lupus erythematosus (SLE): Pleuropulmonary Manifestations

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Agenda

• Introduction
• Pleural involvement
• Pulmonary involvement
• Airway involvement
• Pulmonary vascular disease
• Infections
• Patient Presentation
• Summary
• References
• Acknowledgements
SLE: Epidemiology

• Systemic lupus erythematousus (SLE):
  – Is a chronic autoimmune disease characterized by microvascular inflammation with the generation of autoantibodies that can affect almost any organ system.
  – Approximately 250,000 Americans have systemic lupus. (National Arthritis Data Working Group).
  – Its presentation and course are highly variable.
SLE: Pleuropulmonary involvement

• The majority of patients with SLE develop pleural or pulmonary disease in the course of their illness, diagnosed clinically and/or by images techniques.

• Respiratory involvement is more common in men than in women.

• The pleura is the most common thoracic localization of SLE.
SLE: Pleuropulmonary manifestations

- Infections
- Pleuritis with or without effusion
- Upper and lower airways disease
- Acute lupus pneumonitis
- Alveolar hemorrhage
- Organizing pneumonia
- Chronic interstitial lung disease
  - Lymphocytic interstitial pneumonia
  - Nonspecific interstitial pneumonia
  - Usual interstitial pneumonia
  - Desquamative interstitial pneumonia
- Respiratory muscle weakness (Shrinking lung syndrome)
- Pulmonary hypertension
- Pulmonary embolism
- Mediastinal lymphadenophaty

Crestani B. The respiratory system in connective tissue disorders. *Allergy* 2005; 60: 715-34.
Pleural involvement

- Pleural involvement may be asymptomatic, although pleuritic pain is very common.
- Clinically apparent effusions have been reported in up to 50% of patients and pathological involvement in autopsy in up to 93% of patients.
- Typically associated with chest pain, dyspnea, cough and fever.

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Acute lupus pneumonitis

• Lung parenchyma involvement can be acute or chronic.
  – **Acute lupus pneumonitis:** Non specific, may simulate infection, pulmonary embolism or other. Variable degree of respiratory impairment accompanied by focal or diffuse pulmonary consolidation.

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Acute pulmonary hemorrhage

- **Acute pulmonary hemorrhage:**
  - Bilateral lung infiltrates, ranging from limited ground glass opacities to dense consolidation.
  - Consolidation can be diffuse or patchy.

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Pulmonary fibrosis

• Chronic interstitial pulmonary disease:
  – Pulmonary fibrosis: 
    honeycomb changes with peripheral and basal predominance, linear thickened interlobular septae, ground glass attenuation and parenchymal bands.

Lymphocytic interstitial pneumonia

• **Lymphocytic interstitial pneumonia (LIP):**
  - Usually associated with Sjogren syndrome.
  - Ground glass opacity, poorly defined centrilobular nodules, thickening of bronchovascular bundle, interlobular septae and cystic airspaces.
  - Patchy alveolar infiltrates.

Airway Involvement

• Uncommon in SLE.
• Upper airway, glottis and cricoarytenoid joints seem to be the most commonly involved sites.
Pulmonary vascular disease

- Pulmonary Hypertension:
  - Present in 5-14% of patients. Prevalence tend to increase with time.
  - Few cases result in right heart failure.
  - Diagnosis suspected on echocardiography and confirmed by cardiac catheterization.

Image from PACS, BIDMC
Pulmonary Hypertension

- Dilated main pulmonary artery, abnormalities in perfusion, heterogeneity of lung attenuation (mosaic perfusion).

Images from PACS, BIDMC
Pulmonary embolism

- Pulmonary embolism:
  - Mostly associated with Antiphospholipid syndrome.
  - Chronic pulmonary embolism can lead to pulmonary hypertension.

Pulmonary infections

• Infection is a major cause of morbidity and mortality in SLE.
• 50 % of deaths reported in some series.
• Secondary to immunosuppression associated with SLE itself and induced by corticosteroids and immunosuppressants.
• Susceptible to usual pathogens and opportunistic pathogens.
• Mycobacterial and Nocardial infections seem particularly important
Mycobacterial and Nocardial infection

Mycobacterial Infection: Consolidation and cavitation of left apex.

Nocardial infection: Consolidation and cavitation of right upper lobe.

Shrinking lung syndrome

- Dyspnea associated with small lung volumes, elevated hemidiaphragms and bibasilar atelectasis.
- Syndrome attributed to diaphragmatic dysfunction (myopathy) on the basis of demonstration of decreased respiratory muscle strength.
Shrinking lung syndrome: Images

- Elevated hemidiaphragms, small lung volumes, and bibasilar atelectasis

Our patient

• Our patient:
• 32 years old female.
• Past medical history:
  – SLE course complicated by nephritis, anemia, serositis and ascites.
  – Vascular stenosis resulting in facial edema and subclavian steal.
  – Stage IV sacral decubitus ulcer complicated by osteomyelitis
  – Gastroesophageal reflux disease
Clinical Case: Past medical history

• Past medical history
  – ESRD status post failed renal transplant requiring explant, currently on hemodialysis three times a week.
  – Multiple hospitalizations for line sepsis.
  – History of MSSA endocarditis complicated by embolic stroke and resultant seizure disorder.
  – Sickle cell trait
  – Pulmonary hypertension
  – Restrictive lung disease
Clinical Case: Actual history

• Consults because of history of worsening shortness of breath, worse when lying down in last few weeks. Patient denies any chest pain or cough.

• Findings on physical exam:
  – Febrile up to 101
  – Heart rate: 83, Blood pressure: 110/75, Respiratory rate: 16, SpO2: 98% on 2 liters of O2 on admission
  – Pulmonary exam: Crackles at bases bilaterally no increased work of breathing.
Clinical Case: Laboratory exams

• Findings on laboratory exams:
  – Hematocrit: 23.2%, repeated: 21.8%
  – Hemoglobin: 7.1 mg/dL.

• Patient was transfused one unit of packed red blood cells, with appropriate rise in hematocrit and improvement in shortness of breath.
Clinical Case: Images

- During work up, concerning for multifocal pneumonia vs. pulmonary congestion Chest X Ray (CXR) and Computed Tomography (CT) are obtained...
CXR and Coronal CT

Image from PACS, BIDMC
Chest X Ray

- Findings concerning for multifocal pneumonia though a component of vascular congestion cannot be entirely excluded.
- Increased ground glass opacities.
- Atelectasis and pleural effusions.
Chest CT: Effusions, atelectasis

- Bilateral moderate sized **pleural effusions** worsen since three months ago.
- Adjacent **atelectasis** at lung bases
- Increased heterogeneous air space disease compatible with **multifocal pneumonia**.
Chest CT: Pulmonary edema

- Significant prominence of pulmonary vasculature with septal thickening and patchy ground glass opacity bilaterally compatible with moderate to severe pulmonary edema.
Chest CT: Pulmonary hypertension

- Massively dilated main pulmonary artery (47mm), enlarged azygos vein, compatible with pulmonary hypertension.
Chest CT: Cardiomegaly

- Cardiomegaly with enlarged right atrium and septum thickening.
Summary

• SLE is chronic autoimmune disease that can affect almost any organ system.
• Majority of patients with SLE develop pleural or pulmonary disease in the course of their illness.
• Most common pleuropulmonary manifestations of SLE are:
  – Infections
  – Pleuritis with or without effusion
  – Acute lupus pneumonitis
  – Pulmonary embolism that can lead to pulmonary hypertension
References

• Crestani B. The respiratory system in connective tissue disorders. Allergy, 2005; 60: 715-34
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