Systemic lupus erythematosus and the lung: A pictorial review

Poster No.: P-0090
Congress: ESTI 2015
Type: Educational Poster
Authors: A. Carvalho, P. Leitão, M. S. C. Rodrigues, B. M. Araujo, N. P. Silva; Porto/PT
Keywords: Connective tissue disorders, Diagnostic procedure, CT-High Resolution, CT, Conventional radiography, Thorax, Lung
DOI: 10.1594/esti2015/P-0090

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

To review and illustrate the spectrum of thoracic manifestations in systemic lupus erythematosus.

Background

Systemic lupus erythematosus (SLE) is a chronic, debilitating, autoimmune disease of unknown etiology with multisystem manifestations and a wide variety of presentations. Its severity can range from a benign relapsing-remitting course to a rapidly progressive and not infrequently fatal outcome. SLE is predominantly a disease of women of childbearing age, with peak incidence between the second to fourth decades of life. In adults, females are affected 9 to 13 times more frequently than men. Clinical diagnosis of SLE is made when at least four out of eleven criteria are met (malar rash, discoid lesions, photosensitivity, oral ulcers, nonerosive arthritis, serositis, renal involvement, seizures or psychosis, hematologic abnormalities, immunologic abnormalities and positive antinuclear antibody test).

Thoracic manifestations of SLE are relatively common and over 50% of patients develop pleural or pulmonary disease.

Imaging findings OR Procedure details

PLEURAL AND PERICARDIAL DISEASE (Fig. 1 on page 5, Fig. 2 on page 6)

Pleural disease is the most common thoracic abnormality present in lupus patients:

- **Pleural effusion** or pleural **thickening** occurs in about 70% of cases.
- Pleural effusions in SLE are usually **bilateral, small and exudative**.
- Although isolated pleural effusion is a nonspecific radiologic finding, its presence, particularly when chronic, may suggest the diagnosis of SLE if other clinical features of autoimmune disease are present.
- **Fibrothorax** is a rare complication occurring after refractory pleuritis resulting in thickening of visceral and parietal pleura and usually presents with loculated pleural effusion, pleural thickening and atelectasis of the adjacent lung parenchyma.
Exudative pericardial effusions and pericarditis may occur in up to 50% of patients. SLE can also involve the myocardium and the cardiac valves.

PARENCHYMAL DISEASE (Fig. 3 on page 7, Fig. 4 on page 8, Fig. 5 on page 9)

PULMONARY INFECTIONS

- Pulmonary infections constitute a predictable complication related to immunosuppression by SLE itself or induced by steroid or immunosuppressive therapy.
- Constitutes a major cause of morbidity and mortality (57% of all fatal infections).
- **Pneumonia** is the most common cause of pulmonary infiltrates in SLE patients.
- Although community-acquired **bacterial pneumonia** is the most common cause of pulmonary infection, they may also be due to atypical microorganisms (*Mycobacteria, Pneumocystis jiroveci, Cytomegalovirus, Aspergillus and Nocardia*).
- For patients who are undergoing treatment with high-dose corticosteroids or immunosuppressive drugs, regular screening for **pulmonary tuberculosis** is mandatory in geographic areas in which this entity is prevalent.

ACUTE LUPUS PNEUMONITIS AND DIFFUSE ALVEOLAR HEMORRHAGE (Fig. 6 on page 9, Fig. 7 on page 9)

Acute lupus pneumonitis and diffuse alveolar hemorrhage are less common but serious complications of SLE.

Acute lupus pneumonitis

- Refers to the acute onset of clinical signs and symptoms that may include fever, dyspnea, cough, pleuritic pain and hypoxemia, which is a similar clinical presentation to bronchopneumonia, except that no causative pathogen can be isolated.
- It can occur in up to 12% of patients with a high short-term mortality.
• Usually manifests as patchy unilateral or bilateral areas of **consolidation** or **ground-glass opacity with lower lobe predominance**. Pleural effusions can be present in half of cases.

• Since pneumonia is far more common than acute pneumonitis, it is imperative to exclude an active infection, based on clinical and laboratory findings, before considering this diagnosis.

**Diffuse alveolar hemorrhage**

• Is likely a manifestation of lupus pneumonitis. It manifests as **hemoptysis** (not invariably present), diffuse **alveolar infiltrates** and a **drop in hematocrit** level.

• Usually associated with lupus nephritis, constituting a pulmonary-renal syndrome.

• Consolidation is typically replaced by interstitial abnormalities during resolution.

**SHRINKING LUNG SYNDROME**

This syndrome refers to progressive loss of lung volumes and is thought to be due to diaphragmatic dysfunction or pleuritic chest pain with restriction of respiration.

• Presents with **hemidiaphragmatic elevation**, often with radiologic evidence of linear bibasilar atelectasis and ill-defined juxtadiaphragmatic areas of increased opacity (**Fig. 8 on page 10**).

• Diaphragmatic dysfunction should be suspected when dyspnea is out of proportion to the severity of radiographic abnormalities and when a **restrictive pattern** is shown on pulmonary function tests.

**INTERSTITIAL LUNG DISEASE**

Unlike most connective tissue disorders, chronic interstitial lung disease is a rare manifestation of SLE (estimated prevalence of 3%).

• The most common pattern seen in lupus patients is **non-specific interstitial pneumonia** (NSIP).

• There is a tendency for fibrosis to be **finer** in SLE than in Idiopathic Pulmonary Fibrosis and **honeycombing is very uncommon**.
• Fibrosis predominates at lung bases and lung periphery, although the anterior upper lung zones are also commonly involved. It also tends to be patchy rather than concentric (Fig. 9 on page 11).

**VASCULAR DISEASE (Fig. 10 on page 11, Fig. 11 on page 12, Fig. 12 on page 13, Fig. 13 on page 13)**

• Primary or secondary **pulmonary arterial hypertension** (PAH) is not frequent in SLE (up to 6%).

• The pathophysiology of PAH in systemic lupus is multifactorial and may encompass pulmonary vasculopathy, chronic thromboembolic disease (related to antiphospholipid antibody syndrome), left heart disease, lung disease or the combination of these factors.

• Contrast-enhanced CT may demonstrate the presence of large central pulmonary arteries with pruned peripheral arterial trees and myocardial hypertrophy.

• Contrast-enhanced CT may be warranted when a lupus patient presents with acute dyspnea, because antiphospholipid antibody syndrome acts as an independent risk factor for the development of **deep venous thrombosis** and pulmonary embolism.

**MALIGNANCY (Fig. 14 on page 14, Fig. 15 on page 14)**

There appears to be an increase in malignancy rate in lupus patients compared to the general population with **lung carcinoma** and **lymphoma** being the most common cancers find in this patient group.

**Images for this section:**
**Fig. 1:** Small right pleural effusion. Erect PA chest from this 22 year-old male patient with newly diagnosis SLE shows subtle obliteration of the right costophrenic angle (arrow) consistent with small pleural effusion. The patient had rapidly progressive renal failure from lupus nephritis.
Fig. 2: 65 year-old male patient who presented with dyspnea on exertion and severe anemia. An enlarged cardiac silhouette and left loculated pleural effusion were noted at admission (a). Chest CT (mediastinal window) some days later showed bilateral pleural effusion (pl) and moderate-volume pericardial effusion (per) - panels b and c. The patient had positive ANA test and his anti-dsDNA titer was > 800 U/L. Troponins were elevated, but no chest pain was reported. Acute lupus myopericarditis was diagnosed and treated with pulse corticosteroids and IV cyclophosphamide. PA chest film 2 months later (d) revealed an almost normal-sized cardiac silhouette and no pleural effusion was noted. A discrete linear atelectasis in the left lung base was present. Some pleural plaques related to previous asbestos exposure were also noted.
Fig. 3: Community-acquired bacterial pneumonia in a 56 year-old female with longstanding SLE. There is diffuse infiltrate of the lower half of the right lung on the chest film (a). Chest CT (b) confirmed consolidation of the right lower lobe.
**Fig. 4:** Pulmonary TB. Posteroanterior and lateral chest films (panels a and b) show a micronodular/milliary pattern in both lung fields. In this immunosuppressed male patient with SLE, pulmonary tuberculosis was suspected from clinical and laboratorial findings. HRCT of the thorax (panels c and d) confirmed a milliary pattern consistent with TB. No cavitation or pleural effusion was noted.

![Fig. 4](image1.png)

**Fig. 5:** Pulmonary aspergillosis. HRCT (panel a) shows ill-defined bilateral foci of consolidation and ground-glass attenuation. On the right lower lobe, a soft tissue mass with a surrounding crescent of air was noted (panel b). Bronchoalveolar lavage was positive for Aspergillus fumigatus in this severely immunocompromised patient with SLE and antiphospholipid syndrome. One year later (panel c) there is a well defined cavity with soft tissue mass in the dependent position surrounded by a crescent of air, typical of aspergilloma (arrow).

![Fig. 5](image2.png)

**Fig. 6:** Acute lupus pneumonitis with diffuse alveolar hemorrhage. Anteroposterior chest film (a) shows diffuse opacity of the left lung field and multiple ill-defined opacities on the right lung. The patient was a 72 year-old female with a longstanding history of SLE that presented with hemoptysis and no evidence of infection was found. At CT (panels b and c) diffuse foci of consolidation with basal predominance was reported. These findings are nonspecific and in this case the clinical picture was key to the diagnosis. Bilateral small pleural effusions were also present.

![Fig. 6](image3.png)
Fig. 7: Acute lupus pneumonitis with probable diffuse alveolar hemorrhage. Anteroposterior chest film (a) shows diffuse opacity of both lung fields. At CT (panels b, c and d) there are bilateral medium volume pleural effusions and passive atelectasis of the lower lobes, thickening of the interlobular septa and diffuse ground-glass opacities in both lungs. There was no clinical or laboratorial evidence of infection. These findings are compatible with lupus pneumonitis with possible areas of alveolar hemorrhage although there was no history of hemoptysis.
**Fig. 8**: Diaphragmatic dysfunction. Erect chest PA (a) film demonstrates mild elevation of the left hemidiaphragm, a finding suggestive of diaphragmatic dysfunction in this male patient with SLE. Five years later (panel b), the findings are more expressive, suggesting progressive volume loss. Linear atelectasis at the left lung base (not present before) was also noted.

![Image of chest PA films](image)

**Fig. 9**: Lung fibrosis in SLE. Axial HRCT images in a 30 year-old female patient with SLE showing mild reticular opacities most notable at the periphery of the lung bases. Note the subtle subpleural reticulation at the anterior left upper lobe (arrow).

![Image of axial HRCT images](image)
Fig. 10: Axial contrast-enhanced CT image (mediastinal window) in a female patient with SLE showing an enlarged pulmonary trunk (32 mm), suggesting pulmonary arterial hypertension.
**Fig. 11:** Acute and chronic pulmonary embolism in SLE. Contrast-enhanced axial CT image of the chest in a 56 year-old male patient with SLE who presented with acute dyspnea. There is a small central thrombus (arrow) in the right interlobar artery, favouring acute pulmonary embolism. In panel b, there is a small excentrically located thrombus (arrow) in the interlobar artery. Segmental arteries were normally opacified distally (not shown). This is suggestive of chronic pulmonary embolism.

**Fig. 12:** Massive chronic pulmonary embolism in a 39 year-old male patient with SLE and antiphospholipid syndrome. Contrast enhanced axial CT image (panel a) and maximum intensity projection (MIP) reconstruction (panel b) show massive totally occlusive thrombus (asterisk) with gross calcification at the emergency of the right pulmonary artery. The pulmonary trunk is enlarged (40 mm), reflecting pulmonary arterial hypertension. The patient had recent surgical thrombembolectomy and endarterectomy with restoration of blood flow through the right pulmonary artery (panel c).
**Fig. 13:** Axial HRCT images of the same patient of Fig. 12 demonstrating diffuse mosaic perfusion pattern throughout both lungs, being more expressive on the left. These changes could be due to small airways disease (which is not frequent in lupus patients), however, in this particular case, they were attributed to vascular changes in lung parenchyma (chronic pulmonary embolism).

**Fig. 14:** This 37 year-old female patient with history of SLE presented with dyspnea and weight loss. Chest radiograph at admission (a) showed multiple bilateral lung nodules. A left para-hilar mass was also present. Axial HRCT image (b) depicts bilateral random nodules through both lungs, favoring diffuse metastization. The left para-hilar mass (asterisk) was a biopsy proven lung adenocarcinoma. Two months later, HRCT (panel c) shows disease progression with encasement of the main pulmonary vessels (not shown). The patient died a few days after the scan.
Fig. 15: Lymphoma. Coronal abdominal CT image depicts massive splenomegaly (23 cm in length) in a 52 year-old female with a history of SLE. This was biopsy proven marginal zone non-Hodgkin’s lymphoma.
Conclusion

SLE is an unusually complex multisystem disease and thoracic manifestations of this entity are protean and range from mild, asymptomatic findings to life-threatening events. Radiologists should be aware that pleuropulmonary involvement in this disorder is common and should be able to recognize the plethora of radiographic and CT findings associated with this disease.

References


Personal Information