Pleural and pulmonary involvement in systemic lupus erythematosus

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Summary

Systemic lupus erythematosus (SLE) is a rare complex autoimmune disease with a multisystem involvement. The clinical manifestations of this disease include an erythematous rash, oral ulcers, polyarthritis, nonerosive arthritis, polyserositis, hematologic, renal, neurologic, pulmonary and cardiac abnormalities. The involvement of the respiratory system is frequent. Pleuro-pulmonary manifestations are present in almost half of the patients during the disease course and may be the presenting symptoms in 4–5% of patients with SLE. Complications directly associated to the disease include pleuritis with or without pleural effusion, alveolitis, interstitial lung disease, lupus pneumonitis, pulmonary hemorrhage, pulmonary arterial hypertension, and pulmonary thromboembolic disease. Complications due to secondary causes include pleuro-pulmonary manifestations of cardiac and renal failure, atelectasis due to diaphragmatic dysfunction, opportunistic pneumonia, and drug toxicity. The prevalence, clinical presentation, prognosis and response to treatment vary, depending on the pattern of involvement. As with other connective tissue diseases, early and specific therapeutic intervention may be indicated for many of these pleuro-pulmonary manifestations.

Systemic lupus erythematosus (SLE) is a rare complex autoimmune disease of unknown etiology in which tissues and cells are damaged by pathogenic autoantibodies and immune complexes. SLE most commonly affects women of childbearing age [1]. Women are afflicted more often than men; although when the disease is diagnosed later, there is a lower female/male ratio, less active disease, but greater accumulated organ damage and higher mortality [2].
The most common clinical manifestations of this disease include fever, an erythematous rash, anemia, oral ulcers, polyarthralgia, nonerosive arthritis, polymyositis, thrombocytopenia, renal, neurologic, pulmonary and cardiac abnormalities. Box 1 shows the criteria for the classification of SLE revised by the American College of Rheumatology [3]. The respiratory system is frequently involved by the disease. Pulmonary manifestations may be the presenting symptoms in 4–5% of patients and lungs are involved in almost half of the patients during the disease course [4]. Respiratory involvement is more common in men than in women. All components of the respiratory system may be affected including airways, vessels, parenchyma, pleura and respiratory muscles. Pleuropulmonary involvement of the respiratory system in patients with SLE can be due to complications associated to the disease (primary causes) or concomitant complications (secondary causes) and includes acute or chronic diseases (Box 2). Acute pulmonary involvement tends to develop in association with generalized lupus activity, whereas chronic pulmonary disease may progress independently of disease activity in other organs.

**Pleural involvement**

Pleural disease represents the most common intrathoracic manifestation of SLE, with pathologic evidence of pleuritis or pleural fibrosis in 50–83% of patients at autopsy and pleural effusion in 16–50% on chest radiographs. Pleural involvement may be the first manifestation of SLE and is commonly associated with pericarditis [5]. Clinically lupus pleuritis is characterized by chest pain, dyspnea, cough and possibly fever. Pleuritic pain can be present without radiographically detectable chest effusion. The pleural effusion may be uni- or bilateral, usually small to moderate in size but may occasionally be massive. The pathogenesis of pleural effusion is thought to be due to circulation derived immune complex deposition. Pleural fluid in SLE is a serous or serosanguineous exudate, characterized by a WBC count of 3000–5000 cells/ml with leukocyte differential count showing a predominance of neutrophils or mononuclear cells, near serum levels of glucose, decreased levels of complement and a positive ANA, although none of the immunologic abnormalities described are specific for SLE. Any-way diagnostic thoracentesis is always recommended in an SLE patient with pleural effusion as patients with SLE may have effusions for many different reasons including infection, pulmonary embolism, renal failure, and cardiac failure [5]. At times pleural biopsy is necessary to exclude other etiologies, such as tuberculosis or cancer. Usually lupus pleuritis is very responsive to small doses of systemic corticosteroids, providing a rapid relief of symptoms within days, although spontaneous resolution of SLE effusions may also occur. Chest drainage is necessary in patients with massive effusions, pleural masses, or intractable pleuritic pain.

**Box 1**

**Classification Criteria for the Diagnosis of Systemic Lupus Erythematosus (SLE)**

- **1 Malar rash:** Fixed erythema, flat or raised, over the malar eminences
- **2 Discoid rash:** Erythematous circular raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur
- **3 Photosensitivity:** Exposure to ultraviolet light causes rash
- **4 Oral ulcers:** Includes oral and nasopharyngeal ulcers, observed by physician
- **5 Arthritis:** Nonerosive arthritis of two or more peripheral joints, with tenderness, swelling, or effusion
- **6 Serositis:** Pleuritis or pericarditis documented by ECG or rub or evidence of effusion
- **7 Renal disorder:** Proteinuria >0.5 g/d or 3+, or cellular casts
- **8 Neurologic disorder:** Seizures or psychosis without other causes
- **9 Hematologic disorder:** Hemolytic anemia or leukopenia (<4000/L) or lymphopenia (<1500/L) or thrombocytopenia (<100,000/L) in the absence of offending drugs
- **10 Immunologic disorder:** Anti-dsDNA, anti-Sm, and/or antiphospholipid
- **11 Antinuclear antibodies:** An abnormal titer of ANA by immunofluorescence or an equivalent assay at any point in time in the absence of drugs known to induce ANAs

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rarely needed; unusually, pleurodesis or pleurectomy are necessary for chronic effusions not controlled by steroids [5].

**Pulmonary infections**

Patients with SLE are more susceptible to bacterial and opportunistic infections due to immunosuppressive therapy with glucocorticoids or an immunomodulatory agent as well as characteristic immunologic dysfunction including immunoglobulin deficiency, acquired and inherited complement deficiencies, defects in chemotaxis and phagocytosis and functional asplenia which may account for the increased susceptibility to pneumococcal and Salmonella sepsis [6–9]. Underlying parenchymal disease, atelectasis and respiratory muscle weakness also predispose SLE patients to respiratory tract infections due to bronchial stasis and poor clearance of secretions. The risk of pulmonary infection is three times higher in patients with SLE than in the general population [6]. As infections are among the most important causes of morbidity and mortality in patients with SLE, an aggressive approach to SLE patients with new pulmonary infiltrates is mandatory. Infection should be presumed and treated empirically in any patient presenting with new respiratory symptoms or radiologic imaging until an alternative diagnosis is given [10,11]. In particular, the appearance of focal consolidation makes imperative to exclude an infectious source with both clinical and laboratory evaluation before considering other manifestations such as lupus pneumonitis. Patients with SLE are susceptible to both usual pathogens and opportunistic pathogens. They are prone to bacterial, viral, fungal and protozoan infection. To prevent potentially fatal pneumococcal infections routine pneumococcal vaccination is recommended [12]. Most commonly seen organisms, which cause opportunistic infections are Aspergillus, Cryptococcus, Pneumocystis jiroveci and Cytomegalovirus. Co-infection by both Cytomegalovirus and Pneumocystis jiroveci had been reported and was associated with poor prognosis [13]. Moreover a high prevalence of pulmonary tuberculosis and Nocardia in patients with SLE has been documented [14]. Prednisone treatment, even at moderate doses, and extension of lung involvement have been shown to significantly increase the risk of both usual and opportunistic infections [15–18], whilst antimalarials may have a protective effect [19]. When an infection is suspected bronchoscopic lung sampling should be performed, especially if the patient is receiving immunosuppressive drugs. Recurrent pulmonary infections can lead to bronchiectasis and chronic functional respiratory impairment.

**Acute pulmonary diseases**

Acute lupus pneumonitis (ALP) and diffuse alveolar hemorrhage (DAH) are uncommon, acute life-threatening syndromes associated with SLE resulting from acute injury to the alveolar-capillary unit [20]. ALP occurs in 1–4% of patients with SLE. It can reveal a previously unknown SLE in up to 50% of patients or may occur in the course of the disease. The clinical presentation of ALP is non specific and is characterized by the sudden onset of fever, cough and dyspnea with hypoxemia and hypocapnia, pleuritic chest pain, and patchy alveolar infiltrates on chest radiography, without clinical and laboratory evidence of an underlying infection. Sporadically, hemoptysis may be present. Occasionally, acute respiratory failure, requiring mechanical ventilation will occur. ALP is characterised by diffuse alveolar damage (DAD) microscopically (Figure 1) with alveolar wall...

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**Figure 1**

Autopsy in a 50-year old woman with lupus and acute respiratory failure, showing diffuse alveolar damage (DAD) with fibrin and hyaline membranes (Hematoxylin-eosin, 100 ×). (For courtesy of Dr. Alberto Cavazza).

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**Box 2**

Respiratory system involvement in systemic lupus erythematosus

**Pleural disease**
- Pleuritis with or without pleural effusion

**Parenchymal disease**
- Acute lupus pneumonitis
- Diffuse alveolar hemorrhage
- Interstitial lung disease

**Vascular disease**
- Pulmonary embolism
- Pulmonary hypertension

**Respiratory muscle disease**
- Shrinking lung syndrome
- Inflammatory myopathies
- Drug toxicity
damage and necrosis, alveolar edema, hyaline membranes, inflammatory cell infiltration and alveolar hemorrhage; capillary inflammation and thrombosis are also detected; deposits of immunoglobulins and complement are variably present [21,22]. But these pathologic findings are neither diagnostic nor pathognomonic of ALP.

DAH is a rare but one of the most dreadful manifestation of SLE. It has a prevalence that ranges from 0.5 to 5.7% with a female to male ratio of approximately 6 to 1 [23–31]. DAH is the first manifestation of SLE in 11–20% of the cases. Patients with active renal disease are at increased risk of developing DAH with renal involvement being observed in 60–93% of the patients at diagnosis of DAH [32]. The typical clinical manifestations of DAH are hemoptysis, a rapid fall in the hemoglobin level, and new alveolar or interstitial infiltrates. Fever, dyspnea and cough are also common in these patients. Anemia is usually due to acute blood loss, whereas the extent of hemoptysis is variable and not directly related to the severity of hemorrhage as blood in the acini may not communicate with proximal airways, even in severe cases. Mucocutaneous manifestations, central nervous system involvement, hematologic abnormalities, elevated anti-dsDNA antibodies and low C3 and C4 complements are often observed in these patients [23–31].

Arterial hypoxemia is common and more than 50% of the patients will need mechanical ventilation. The pathogenesis of the disease is not completely understood. A proposed mechanism is immune-mediated damage of small blood vessels and alveolar septae [32–34]. Actually, deposition of IgG and C3 in the capillary basement membrane, alveolar walls and blood vessels of involved lung tissue have been demonstrated by immunofluorescence and electron microscopy [32,33]. Different pathological conditions related to infections such as disseminated Strongyloides and Cytomegalovirus resulting in immunoglobulin and complement deposition may also cause membrane damage and pulmonary hemorrhage [34,35]. Other possible mechanisms include the presence of vasculitis, anti-phospholipid antibodies, and anti-basement membrane antibodies [36,37]. Histopathologic findings of DAH are nonspecific (Figure 3). Most cases are associated with bland hemorrhage without changes of capillaritis or any associated interstitial inflammation [30,38]. Diffuse injury to the microvasculature known as capillaritis with neutrophil infiltration of alveolar septae often associated with destruction of the alveolar wall has also been described [23,31,37]. But capillaritis is not specific of SLE as it has also been described in alveolar hemorrhage associated with the antiphospholipid syndrome, in polymyositis (PM) and other connective tissue diseases (CTD), in Henoch–Schoenlein purpura, cryoglobulinemia, Behcet syndrome, and Wegener disease [39–41]. It may also be seen in interstitial and intra-alveolar macrophages (hematoxylin-eosin, 40 ×). B. Higher magnification of Figure 2A showing blood and fibrin filling the alveoli (Hematoxylin-eosin, 200 ×) (for courtesy of Dr. Alberto Cavazza).

ALP can be difficult to differentiate from infectious pneumonia because of similar clinical and radiological presentation. Although ALP and DAH are histopathologically distinct, the imaging features are similar and a radiological differentiation between the two entities can be difficult. Bilateral patchy airspace opacification due to consolidation or ground glass opacities which usually predominate in the lower lobes are found in both conditions [42]. Rapid resolution of radiological
changes may favour pulmonary haemorrhage, whereas ALP may be associated to pleural effusions. Patients with DAH may also present with unilateral infiltrates but these are uncommon. However, even in the presence of extensive hemorrhage, chest radiographs may be normal [29]. Evaluation of carbon monoxide diffusing capacity (DLCO) can help in the diagnosis of early hemorrhage. An increase in DLCO of 30% or more over the baseline level or an elevation of 130% or more of the predictive value is suggestive of pulmonary hemorrhage [43] (Figure 3). When acute pulmonary disease in SLE patients is suspected early diagnosis is vital since prompt therapeutic interventions are crucial for survival. Bronchoalveolar lavage is the most relevant tool for the diagnosis of DAH and must be always performed, if clinically feasible. A search for common and opportunistic agents is also necessary. The presence of blood and haemosiderin-laden macrophage in bronchoalveolar lavage with the absence of purulent sputum and organisms favors the diagnosis of pulmonary haemorrhage. Concomitant lung infection is observed in about 30% of patients with DAD, and bears a poor prognosis [22]. Although lung biopsy (transbronchial or open) can aid to establish a definite diagnosis of lupus pneumonitis or DAH, it is associated with a high morbidity and may be dangerous in critically ill patients, thus it is not recommended unless in selected cases. As there are no controlled studies for the treatment of ALP and DAH, the current recommendations are based on case reports and clinical

**Figure 3**

*Acute lupus pneumonitis in a young woman.*

A, B, C, D: Acute respiratory failure required an admission in intensive care unit. Computed tomography of the lungs showed bilateral ground glass opacities and consolidations.
experience. Corticosteroids are generally accepted as the first line of therapy for acute immune-mediated lung injury in patients with SLE. Preferred treatment for ALP is pulse methylprednisolone (250–1000 mg/day for several days) in patients with severe initial presentation. Immunosuppressive or cytotoxic agents such as azathioprine or cyclophosphamide may be associated in patients with a poor response to steroids [14,44]. Pulse methylprednisolone therapy (1000 mg daily for 3 days) is used for DAH. By the fourth day, methylprednisolone dose is decreased to 1–2 mg/kg/day, switching to oral prednisone (1 mg/kg/day) over the next days [25] As high dose or IV methylprednisolone can be associated with higher incidence of serious infections, a lower methylprednisolone dose (500 mg daily) may be used as initial treatment [25]. Reports in the literature suggest that use of cyclophosphamide confers a survival benefit in patients with DAH. One recent case series in which cyclophosphamide was used in the majority of patients showed a high mortality rate (over 50%) [45]. On the other hand, the combination of pulse methylprednisolone and cyclophosphamide has been related to increased survival rates in others studies [28,29]. The efficacy of other immunosuppressive agents such as cyclosporine, azathioprine, and tacrolimus for the treatment of pulmonary hemorrhage is still uncertain [30,46]. Plasmapheresis as an adjunct to high-dose corticosteroids and cyclophosphamide has been used with good outcomes in rapidly deteriorating patients [25,27,30]. Interestingly, a 100% survival rate has been reported when prophylactic antibiotics were added to pulmonary hemorrhage therapy [24]. Thus, these patients should be thoroughly evaluated for coexistent infections and broad spectrum antibiotics instituted without delay. The overall prognosis for ALP is poor with 50–90% mortality despite treatment. Prompt identification and treatment are essential for survival during the acute phase. For those who survived the acute episode, 50–100% would eventually progress to chronic interstitial pneumonitis [47]. DAH carries a poor prognosis with mortality rates of 70–90% and survivors can develop pulmonary fibrosis [29,48].

**Chronic pulmonary diseases**

Diffuse interstitial lung disease (ILD) is relatively less common in SLE than in other CTDs, although it may dominate the clinical picture in some patients. The involvement of the lung affects 3–8% of SLE patients, with a progressive increase in prevalence with disease duration [49,50]. However, different high resolution computed tomography (HRCT) series have demonstrated a higher rate of ILD features (about one third of SLE patients), suggesting that subclinical lung disease is common [51–53], although the majority of these patients had mild interstitial changes with interlobular septal thickening being the most common finding.

As recent reports are scarce, our knowledge on the clinical and imaging patterns of ILD of clinically relevant disease is mostly based on older studies. ILD is more common in older patients, men, and patients with late-onset SLE [50,51]. ILD in SLE may present either as a sequelae of ALP [54,55] or may have a more insidious onset [55,56] with a chronic non productive cough, dyspnoea on exertion and recurrent pleuritic pain. Histological reports show nonspecific abnormalities with interstitial lymphocytic infiltrates, interstitial fibrosis and honeycomb changes [55,56]. *Nonspecific interstitial pneumonia* (NSIP) is likely the most common pattern in SLE patients with pulmonary fibrosis shown by HRCT [57] (Figure 4), although the real incidence of NSIP in SLE is still not well defined [58]. Lymphocytic interstitial pneumonia (LIP) has been also described in a few patients with SLE, usually in association with Sjögren’s syndrome. In these cases, the development of lung cysts should suggest the diagnosis [59–61]. Finally, the clinic-radiological syndrome of organizing pneumonia (OP) with patchy alveolar infiltrates and an histologic pattern of organizing pneumonia has been described in patients with SLE. These cases usually show a good response to corticosteroids [62–64]. The simultaneous occurrence of SLE and sarcoidosis has been observed in a few cases [14]. Nodular amyloidosis and excavating nodules have also been reported.
Pulmonary embolism

SLE patients are at increased risk for coagulation problems. Pulmonary embolism should be suspected in patients who have antiphospholipid antibodies (aPL) with acute respiratory symptoms such as pleuritic chest pain and dyspnoea. Antiphospholipid antibodies are a family of acquired autoantibodies that are associated with vascular thrombosis and may be present in up to two thirds of patients with SLE [69,70]. The two most well known and clinically important are the lupus anticoagulant (LA) and anticardiolipin antibodies (aCL).

The combination of clinically important vascular events and the presence of LA or aCL is known as antiphospholipid syndrome (APS). A metaanalysis found that subjects with aPL and SLE were more than six times more likely than subjects with SLE and no aPL to develop venous thromboembolism [71]. Patients with LA have been shown to have a greater risk than patients with aCL [69,71]. Clinical features of pulmonary embolism depend on the degree of obstruction. With massive pulmonary embolism, right ventricular failure with acute circulatory collapse may be present. Dyspnoea, pleuritic pain, haemoptysis, crepitations and tachypnoea are most common manifestations in less severe cases. Pulmonary hypertension (PHT) is likely to be the consequence of chronic thromboembolism. Nonthrombotic intrathoracic complications associated with the aPL include pulmonary arterial hypertension, DAH, ARDS, and cardiac valvular lesions [72]. The term catastrophic APS (CAPS) refers to a syndrome due to small-vessel occlusion in three or more organs, that may develop in some patients with APS; it is often associated with infections, neoplasm, and surgery. CAPS usually involve the cardiopulmonary system with respiratory failure and rapid progression to ARDS [73,74]. For patients with aPL, the role of prophylactic anticoagulant or antiplatelet therapy is still debated. Controversial is also the treatment of arterial thrombosis, with some experts recommending high-intensity anticoagulation. The use of corticosteroids or immunosuppressive agents as adjuvant should be considered to decrease circulating antibodies. The placement of inferior vena cava filter may be considered in individual cases [75]. The treatment of CAPS includes anticoagulation, glucocorticoids with an immunomodulatory agent, and often plasmapheresis and IV Ig.

Pulmonary arterial hypertension

CTDs such as systemic sclerosis (SSc) and SLE can be complicated by severe pulmonary arterial hypertension (PAH) [76]. Previous studies have largely concentrated on SSc-associated PAH (SSc-PAH), with a disease prevalence estimated to be between 7.5 and 12% [77,78]. Some degree of PHT complicates the course of SLE in 5–14% of the patients [79–81]. PHT prevalence and severity of disease in patients with SLE tend to increase with time [82]. Autopsy findings from patients with SLE and severe PAH have demonstrated alterations of medial hypertrophy, intimal fibrosis, and plexiform lesions, which are virtually identical to the pathologic changes seen in patients with idiopathic PAH [83]. Pulmonary veno-occlusive disease, a rare form of pulmonary hypertension with different histopathology, has also been reported [84]. The pathophysiology of SLE-PAH is poorly understood; antiphospholipid antibodies, anti-endothelial cells antibodies, vasculitis, vasospasm, and inflammation all contribute to the development of the typical proliferative lesions observed in the disease. Raynaud’s phenomenon, antiphospholipid antibodies and ILD are more common in SLE patients with PAH [14].

The diagnosis of PAH is suspected on echocardiography and must be confirmed by right heart catheterization. Exclusion of chronic thromboembolism with ventilation-perfusion scintigraphy is always necessary. In SLE estimated survival has ranged from relatively good to being worse than in idiopathic PAH [85,86]. Treatment is based on oral anticoagulants and vasodilators. Intravenous epoprostenol has been used with good results [84,87]. Newer vasodilators, such as sildenafil and endotelin-antagonists may be useful in some patients. Studies in small collectives of patients with SLE-PAH reported beneficial effects of immunosuppressive therapy, highlighting a possible link between a systemic inflammatory condition and pulmonary vascular disease [88,89]. PAH associated with SLE may respond to a combination of IV bolus of cyclophosphamide and systemic glucocorticoids. Patients who have less severe disease at baseline are those who could benefit from this immunosuppressive therapy while for patients with more severe disease, pulmonary vasodilators should be started, possibly in combination with immunosuppressive treatment [90].

Myopathies and muscle dysfunction

Shrinking lung syndrome (SLS) is a manifestation of SLE characterized by progressive dyspnea, the characteristic chest radiographic findings of small lung volumes, elevated hemidiaphragms and bibasilar atelectasis, with a restrictive ventilatory defect and a preserved carbon monoxide transfer...

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coefficient. The exact prevalence of SLS in patients with SLE is unknown; occurrence has been described in 18 to 27% of patients [91,92]. The pathogenesis of this condition is still not entirely known. It was initially attributed to diaphragmatic dysfunction on the basis of the demonstration of decreased inspiratory muscle strength in 11 SLE patients [93]. On the other hand, Laroche et al. using bilateral electrostimulation in 12 patients with the SLE, failed to demonstrate diaphragm weakness [94]. In a case-report by Hardy et al. a patient with the syndrome and bilateral phrenic nerve paralysis has been described. With corticosteroids, the phrenic nerve function recovered without improvement of the restrictive functional pattern, suggesting that reduced diaphragm muscle contractility per se does not explain the small volume lungs and respiratory symptoms in patients with the syndrome [95]. Patients with SLS refer exertional dyspnea that progresses for weeks or months, leading to a marked reduction in tolerance to exercise. Pleuritic pain is also a frequent symptom in these patients.

Physical examination shows a marked limitation of the thoracic movement and the use of accessory muscles. Lung auscultation is usually normal, although bibasilar rales can be heard as the result of atelectasia. Elevation of a hemidiaphragm and basal atelectasia can be seen on chest X-ray, as well as pleural thickening and pleural effusion. SLS can be present in any phase of the disease or can even be its first manifestation. The prognosis of patients with SLS is usually favorable [96]. Treatment of SLS is still empirical. In most cases, moderate-high doses of steroids are used [91,97,98]. However, complete recovery of vital capacity is not frequent. Teophyllin and beta-agonists have also been shown as useful in SLS [92]. Anecdotal data also supports the use of other immunosuppressants, such as cyclophosphamide or azathioprine in patients unresponsive to steroids [91].

Other causes of myopathy in patients with SLE include inflammatory myopathies, both related to PM and dermatomyositis (which can accompany SLE in up to 10% of cases) as well as myopathy related to the disease. Secondly, myopathy can be due to drug toxicity, including mainly steroids, antimalarials, and statins [99,100]. Other less frequent causes of myopathy in SLE are “myastheniform” diseases or concurrent myasthenia gravis and muscle weakness due to vitamin D, which is normally insufficient in patients with SLE due to the use of sunblock, the lack of sunlight exposure, and the activity of the disease itself [101].

Conclusion

To conclude, pulmonary manifestations of lupus erythematosus are myriad. CT scan has emerged as a very useful tool in the early diagnosis and management of pulmonary diseases associated with SLE. The necessity of ruling out infection and establishing the diagnosis of acute pulmonary involvement such as alveolar haemorrhage, lupus pneumonitis, pulmonary embolism or acute alveolitis in ILD is vital as these conditions need immediate treatment and any delay can result in increased morbidity and mortality.

Conflicts of interests: None.

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PULMONARY INVOLVEMENT IN SYSTEMIC DISEASES


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