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Editorial

Pulmonary Manifestations of Rheumatoid Arthritis

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Rheumatoid arthritis (RA) is a systemic autoimmune process classically known for chronic symmetrical erosive synovitis. It is generally progressive. Lungs are the site of a myriad of non-articular manifestations of RA. Other non-articular manifestations can include subcutaneous nodules, vasculitis, pericarditis, mononeuritis multiplex, and episcleritis.

Pleuropulmonary manifestations of RA include:

1. RA associated interstitial lung disease (ILD)
2. Pulmonary nodules
3. Large and small airway obstruction
4. Pleural disease
5. Vascular disease (including vasculitis and pulmonary hypertension).

In addition, pleuropulmonary infections can occur (related to RA itself as well as drug-induced immunosuppression) and drug-induced pulmonary toxicity can occur related to medication use to treat rheumatoid arthritis. Furthermore, in a given patient, multiple pleuropulmonary syndromes can overlap (for example interstitial lung disease and pleural thickening). Prevalence of RA associated respiratory disease is difficult to estimate because of variations in study population and different techniques utilized to detect disease (HRCT versus pulmonary function tests versus autopsy, etc). In many cases, pleuropulmonary involvement can be subclinical, which further complicates epidemiologic assessment. Overall, the most common pleuropulmonary manifestations of RA appeared to be interstitial lung disease (ILD) and pleural disease.

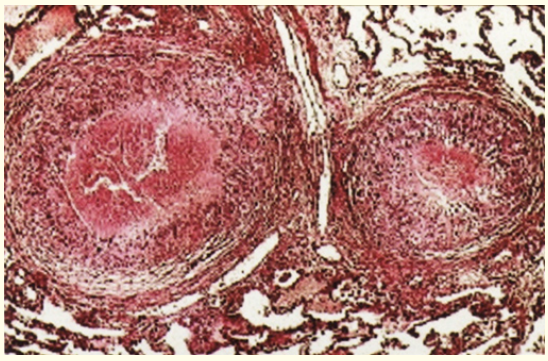
RA related ILD can include various histologic patterns including nonspecific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), organizing pneumonia (OP), lymphocytic interstitial pneumonia (LIP), desquamative interstitial pneumonia (DIP), and acute interstitial pneumonia (AIP). Patients with RA related ILD that are most likely to benefit from aggressive immunosuppression include younger patients, patients with histopathologic patterns other than UIP, and/or evidence of physiologic and/or radiographic progression over the proceeding three to six months.

Pleural disease is most common in patients with longstanding RA, but can precede joint disease. It is more common in men and coexists with rheumatoid nodules and ILD in up to 30% of patients. RA related pleural disease can often be subclinical. It can include exudative inflammatory pleural effusions. Rheumatoid nodules can develop necrosis and cavitation and rupture into the pleural space with creation of a bronchopleural fistula. Other manifestations include chyliform or "cholesterol" pleural effusion as well as "trapped lung." Empyema also needs to be considered in the differential diagnosis of these immunocompromised patients when they present with a pleural effusion.

Upper airway obstruction can occur in RA because of cricoarytenoid arthritis; less common causes include vasculitis involving the recurrent laryngeal or vagus nerves, which can then cause obstruction due to vocal cord paralysis. Upper airway disease is more common in women and in longstanding RA. Unfortunately, symptoms of upper airway obstruction can often be absent until significant airway

obstruction occurs and the patients can present with stridor. It is important to remember that because RA can be complicated by cervical spine instability, intubation should be performed by highly experienced clinicians with care to avoid excessive neck flexion.

Small airway dysfunction is known to occur in up to 24% of non-smokers with RA. Small airway abnormalities can be seen on HRCT in many patients who did not have physiologic evidence of airway obstruction. This phenomenon is of unclear clinical significance. Obliterative bronchiolitis (OB) is a rare (and usually fatal) condition characterized by progressive concentric narrowing of membranous bronchioles. OB has been associated with both RA itself and drugs utilized in the treatment of RA. OB appears to be more common in women.



Follicular bronchiolitis (lymphoid hyperplasia of bronchus associated lymphoid tissue) can also occur in rheumatoid arthritis (either alone or in combination with NSIP). On HRCT, this can cause centrilobular or peribronchial micro-nodules (less than 3 mm) with branching linear structures, which can include bronchial dilation and bronchial wall thickening. Bronchiectasis has also been reported in up to 30% of patients with RA and can occur without evidence of ILD.

Rheumatoid nodules are the only pulmonary manifestations specific for RA. Rheumatoid lung nodules occur more often in patients with a longer duration of disease and concomitant subcutaneous rheumatoid nodules. They are usually located in subpleural areas or interlobular septa, range in size from a few millimeters to several centimeters and may be single or multiple, solid or cavitary. Rheumatoid nodules can cause hypermetabolism on a PET scan (even in the case of nonmalignant rheumatoid nodules). Rheumatoid nodules can resolve spontaneously and complications such as bronchopleural fistula are infrequent.

Caplan's syndrome refers to a combination of RA and occupational dust exposure (pneumoconiosis). This can cause rapid development of multiple basilar nodules with mild airflow obstruction. Caplan's syndrome can also be complicated by the development of progressive massive fibrosis.

Drug-induced lung disease in the setting of RA is beyond the scope of this article, but drug reaction should be considered in the differential diagnosis of physiologic and radiographic abnormalities in patients with RA. RA can also cause thoracic cage abnormalities, which can impact pulmonary function. RA appears to increase the risk of venous thromboembolic disease slightly (even after controlling for other risk factors). There also appears to be a slightly increased risk of developing lung cancer in patients with RA when compared to the general population.

Primary pulmonary vasculitis is quite rare. Pulmonary hypertension can be related to underlying vasculitis. Clinical manifestations can be similar to those of idiopathic pulmonary arterial hypertension. Secondary pulmonary hypertension (WHO Group 3) can occur in the setting of RA related ILD.

Given the multiple pleuropulmonary manifestations of RA, the monitoring and management of these patients can be quite challenging and often will involve close collaboration between a variety of specialists including a pulmonologist and a rheumatologist. Pulmonary infection can be a major contributor to morbidity and mortality in patients with RA. Vaccination against Pneumococcus and influenza should be considered in all patients with RA. Pneumocystis prophylaxis should be considered in some patients with RA (depending on their level of immunosuppressant).

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