Insights into pleuro-pulmonary high-resolution computed tomography manifestations of rheumatoid arthritis

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Learning objectives

- To describe the characteristic pleuro-pulmonary manifestations of rheumatoid arthritis (RA)
- To illustrate the key aspects of lung involvement in RA at high-resolution computed tomography (HRCT);
Background

Rheumatoid arthritis (RA) is a chronic and progressive autoimmune disorder mainly affecting the joints, but also showing a broad spectrum of extra-articular manifestations. Intrathoracic involvement is common and associated with substantial morbidity and mortality (1). RA causes a variety of pleuro-pulmonary lesions, concerning all compartments: pulmonary parenchyma, pleura, small and large airways, pulmonary vasculature and lymphatics (1). HRCT can accurately depict these lung changes, furthermore allowing for an early diagnosis in asymptomatic patients. Likewise the role of HRCT has been well established in the differentiation between nonspecific interstitial pneumonia- NSIP and usual interstitial pneumonia-UIP pattern, this distinction having important clinical implications regarding the management and outcomes of RA patients - the UIP pattern being associated with significantly shortened survival compared to NSIP pattern (2).
Findings and procedure details

The characteristic findings include: interstitial lung disease (ILD), presenting as UIP pattern, NSIP pattern, organizing pneumonia-OP, rarely lymphoid interstitial pneumonia-LIP; airway disease: bronchial wall thickening, bronchiectasis, follicular bronchiolitis, bronchiolitis obliterans; pleural diseases: pleural thickening, pleural effusion and spontaneous pneumothorax; rheumatoid nodules (3). Pulmonary complications are mainly secondary to vascular involvement (pulmonary arterial hypertension- PAH) drug toxicity, infections or exacerbations.

Interstitial lung disease:

Usual interstitial pneumonia: is the most encountered pattern of ILD in RA and this entity has a poorer prognosis than NSIP, OP or LIP (6), independently of other coexisting patterns. These patients typically present slowly progressive dyspnea and nonproductive cough. The major histopathologic feature of UIP is the presence of fibroblastic foci alternating with areas of normal lung or interstitial inflammation and honeycombing. 

HRCT typically reveals: reticular opacities, honeycombing, often associated with traction bronchiectasis plus an apicobasal gradient and peripheral predominance (Fig. 1); ground-glass opacities are frequently present but are limited in extent.

Nonspecific interstitial pneumonia: this type of ILD is a common finding in patients with RA and is associated with a variety of histologic and radiologic appearances, making the diagnostic approach quite difficult. Symptoms are usually nonspecific and similar to those of UIP, but less intense and the response to treatment of these patients is superior, compared with patients that present UIP pattern (7).

HRCT typically reveals: patchy ground-glass opacities combined with irregular linear or reticular opacities and scattered micronodules (8), usually bilateral and with a predominance of subpleural and basal regions (Fig. 2); other HRCT findings may include in cases of advanced disease traction bronchiectasis (Fig. 2), consolidations and subpleural cysts, which is an uncommon finding; however the HRCT hallmark of NSIP remain the extensive ground-glass opacities (6);

OP (organizing pneumonia): is not an unusual manifestation in patients with RA and the histologic hallmark corresponds to bronchiolitis obliterans organizing pneumonia: the presence of granulation tissue polyps in the alveolar ducts and alveoli. Patients typically present respiratory symptoms developed over a few weeks such as: dyspnea, nonproductive cough, fever but the majority recover completely after corticosteroid administration (6).
HRCT typically reveals: multiple patchy airspace consolidations, variable sized (from few centimeters to an entire lobe), usually bilateral, often migratory, with characteristic peripheral or peribronchial distribution in the lower lung lobes (Fig. 3); air bronchogram is commonly seen in these areas; the appearance of the lung opacities can vary from ground-glass to consolidation and the revers halo sign (ground-glass opacity surrounded by a ring of consolidation) is evocative for OP.

Lymphoid interstitial pneumonia: is a benign condition that needs to be differentiated from follicular bronchiolitis due to overlap of clinical and histopathologic patterns. The distribution of infiltrates are diffuse in LIP whereas in follicular bronchiolitis are predominantly peribronchial and centrilobular (6).

HRCT typically reveals: poorly defined centrilobular nodules, areas of ground-glass pacity, interlobular septal thickening, bronchovascular bundle and thin-walled cysts (1-30 mm); moreover, the CT features of LIP are superimposed with those of cellular NSIP such as: frequent lower lobe localization and areas of ground-glass attenuation; the key aspect is the cystic pattern more common and extensive in LIP.

Pleural disease:

Pleural thickening and pleural effusion: are the most encountered thoracic manifestations of RA, found in 38%-73% (4), commonly in middle-aged men with positive rheumatoid factor (4). Most pleural effusions are small, asymptomatic, exudative in nature and resolve spontaneously. Large effusion can cause different symptoms like chest pain, dyspnea and may indicate an underlying lung disease.

HRCT typically reveals: unilateral small or loculated effusion that, in most cases, associates pleural thickening;

Spontaneous pneumothorax: may develop directly from the rupture of a subpleural necrobiotic rheumatoid nodule, or through the formation of a bronchopleural fistula (Fig. 7).

Rheumatoid nodule:

The pulmonary rheumatoid nodules are histologically similar to those found in subcutaneous tissue (necrobiotic nodules that have a central eosinophilic fibrinoid necrosis) (5) and are usually associated with advanced disease.

HRCT typically reveals: multiple well-defined nodules, from few millimeters up to 5 centimeters (4), mostly located in peripheral zones of the upper and middle lung regions (Fig. 5). Occasionally these nodules are seen on ILD background and may undergo some changes over time such as: enlarging, cavitation (Fig. 4) or spontaneous resorption.
Rarely these nodules can rupture into the pleural space, producing pneumothorax, pleural effusion or empyema and these appearances must be distinguished from malignant and infectious lesions.

**Airway disease:**

*Bronchial wall thickening and bronchiectasis:* can be an isolated common finding or much more frequently associated with ILD or bronchiolitis obliterans.

*Bronchiolitis obliterans:* is seldom encountered in RA and is represented by progressive airflow obstruction (concentric fibrous narrowing of the bronchioles) (3).

*HRCT typically reveals:* mosaic attenuation pattern; the air trapping is typically observed on expiratory scans; other finding may include: bronchial wall thickening, bronchiectasis, centrilobular emphysema and centrilobular nodules.

*Follicular bronchiolitis:* is a rare benign condition, histologically characterized by the presence of hyperplastic lymphoid nodules in peribronchial and peribronchiolar areas (3).

*HRCT typically reveals:* small bilateral centrilobular and peribronchial nodules, which often show ground-glass attenuation, bronchial wall thickening, peribronchial thickening and interlobular septal thickening.

**Complications:**

*PHA:* is the main vascular complication of RA which may occur in isolation or in combination with ILD; patients with PAH present an increased diameter of the pulmonary arterial trunk (>29 mm), the main pulmonary arteries and their segments, and, in more advanced cases, the right heart chambers and azygos-hemiazygos venous system (9).

*Drug toxicity:* various drugs may cause lung disease; for example, gold salts and penicillamine may cause diffuse alveolar damage and obliterative bronchiolitis; pneumonitis usually develops during the use of methotrexate; the diagnosis is made by excluding other possible causes, such as infections and exacerbation of ILD (10);

*Infections:* these are secondary processes mainly present in patients undergoing treatment with corticosteroids, disease-modifying agents (DMARDs) and TNF# antagonists; the pulmonary infections are represented by: bacterial pneumonia, pulmonary tuberculosis, pulmonary aspergillosis (Fig. 5 and Fig. 6), pulmonary cryptococcosis, and Pneumocystis jiroveci pneumonia (3).
**Acute interstitial pneumonia**: is a rare and rapidly progressive condition (histologically translated as diffuse alveolar damage), characterized by multiple patchy areas of ground-glass opacities most often associated with areas of focal lung sparing, which produce a geographic appearance (11).
Images for this section:

**Fig. 1:** Image A and B: Axial CT scan of a 62-year-old patient with RA shows: UIP pattern-peripheral reticulation and honeycombing cysts (black arrow) with apicobasal gradient, cylindrical bronchiectasis and bronchial wall thickening (signet ring sign- red arrow)

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**Fig. 2:** Axial HRCT scan (images A and B) and sagittal reformat (image C) of a 59-year-old patient with RA shows: typical aspect of NSIP with reticulation and ground-glass opacities as well as traction bronchiectasis (red arrow); the apicobasal gradient and traction bronchiectasis (red arrow);

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Fig. 3: Axial HRCT scan (image A) and coronal reformat (image B) of a 39-year-old patient with RA show: typical aspect of OP with peripheral bilateral consolidations (red arrows) in the lower lung lobes with air bronchogram;

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**Fig. 4:** Sagittal HRCT reformat of a 73-year-old patient showing rheumatoid cavitating pulmonary nodules (red arrows);

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Fig. 5: Coronal reformat (images A and B) of a 86-year-old patient showing multiple lung rheumatoid nodules, one of them presenting air crescent sign/Monad sign, typical for aspergilloma (red arrow);

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Fig. 6: Axial CT scan of a 61-year-old patient with RA shows: pulmonary nodules with ground-glass halo (red arrow) in a case of angioinvasive aspergillosis;

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**Fig. 7:** Axial CT scan (image A) and coronal and sagittal reformats (image B and C) of a 78-year-old patient with RA shows: multiple pulmonary rheumatoid nodules, one of them showing air crescent sign/Monad sign, typical for aspergilloma (black arrow), and air-fluid level within the pleural cavity, demonstrating a hidropneumothorax (blue arrow); fistula (red arrow) between a rheumatoid cavitating nodule and the pleural space.

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Conclusion

HRCT allows for an accurate characterization of pleuro-pulmonary involvement in RA and has become essential in the accurate identification, early diagnosis, management and prognosis of RA patients.
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References


