The thoracic manifestations of rheumatoid disease - a pictorial review

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Learning objectives

Pleural and pulmonary disease is a frequent extra-articular manifestation of rheumatoid arthritis and is responsible for approximately 20% of rheumatoid associated mortality. We aim to describe the imaging features associated with this using plain films, CT and MRI from a series of patients with rheumatoid arthritis investigated at our institution.

Background

Rheumatoid arthritis is a relatively common multisystem disease associated with significant mortality and morbidity. Thoracic disease, both pleural and pulmonary, is a frequent extra-articular manifestation of rheumatoid arthritis and is responsible for approximately 20% of rheumatoid associated mortality.

Rheumatoid disease and its associated therapies can affect all compartments of the lung inciting a range of stereotyped pathological responses and it is not infrequent for multiple pathologies to co-exist.

In some instances development of pulmonary complications may precede typical rheumatological presentation of the disease and be the first indication of an underlying connective tissue disease. The spectrum of thoracic pathology related to rheumatoid arthritis is reviewed.

Imaging findings OR Procedure details

Pleural Disease

Pleural disease is the most common thoracic feature of rheumatoid disease identified in up to 50% of patients at post-mortem [1] although only 20% of patients are thought to be symptomatic. Effusions are usually unilateral, may be loculated and are frequently associated with subcutaneous nodules and pericardial effusions [2].

Chest radiography remains the initial radiological examination for pleural disease (Fig 1) with a PA radiograph demonstrating effusions of a volume >200mls. Ultrasound
and CT are even more sensitive and can distinguish pleural thickening from effusions (Fig 2) helping to characterise features such as loculation and distribution, as well as highlighting features that might suggest malignancy rather than inflammatory disease. Magnetic resonance imaging (MRI) provides detailed imaging of the pleura (Figs 3, 4) and may be useful in distinguishing benign from malignant disease but is generally used for problem solving rather than first line imaging.

The presence of a chronic effusion may lead to fibrous visceral pleural thickening causing a trapped lung (Fig 5) preventing lung re-expansion despite drainage of the effusion [3].

**Parenchymal Lung disease**

Pulmonary rheumatoid nodules (RN) or necrobiotic nodules are found in 1% of chest X-rays (Fig 6) and in up to 20% of HRCTs (Fig 7). They are usually associated with subcutaneous nodules and appear in the advanced stages of the disease, although the nodules themselves are asymptomatic and do not reflect disease activity [4]. Nodules can range from millimeters to a few centimetres, are generally well defined and peripheral, and cavitate in up to 50% of cases (Fig 8).

Differentiating rheumatoid nodules from lung metastases or primary malignancy can be difficult radiologically and the use of PET-CT is of limited value with both entities potentially showing avid uptake of FDG [5]. Regression with time or during treatment may be helpful in the diagnosis but ultimately biopsy is often necessary.

**Interstitial lung disease**

Rheumatoid arthritis causes clinically important interstitial lung disease (ILD) in around 10% of affected patients and mortality related to RA-ILD is increasing as a proportion of total rheumatoid related deaths [6]. RA-ILD covers a wide range of pathologies: usual interstitial pneumonia (UIP) (Fig 9) and non-specific interstitial pneumonia (NSIP) (Figs 10, 11) are by far the most common patterns of disease but organizing pneumonia (Fig 12), lymphoid interstitial pneumonia (Fig 13) and amyloid can also occur.

Conventional radiographic abnormalities occur in only 1-6% of patients with rheumatoid arthritis but are of limited diagnostic use. HRCT studies detect radiological abnormalities in around one third of patients and allow more confident disease characterisation [7]. Radiographic and HRCT findings for RA-ILD are the same as for the idiopathic interstitial pneumonias but frequently co-exist with other pulmonary pathologies, such as airway or pleural disease, which may be a useful clue to their underlying cause [8].
Airways disease

There is a high prevalence of airways disease in patients with RA which can affect large, medium and small airways.

Airway disease in the form of bronchiectasis (Fig 14) occurs in up to 30% of RA patients [9] although clinically significant disease occurs less frequently. Obliterative (constrictive) bronchiolitis (Figs 15, 16) is also common and may be found with or without bronchiectasis.

An uncommon small airway complication of RA is follicular bronchiolitis (Fig 17), also known as lymphoid pulmonary hyperplasia, which falls in the spectrum of LIP but shows a bronchocentric propensity. It is primarily a histopathologic diagnosis characterised by the presence of hyperplastic lymphoid follicles with reactive germinal centres which may narrow or obliterate the bronchiolar lumen [10].

Drugs

The majority of drugs used in the treatment of RA are associated with pulmonary complications that are frequently indistinguishable, clinically, radiologically and histologically from RA-ILD. HRCT findings are rarely drug specific and reflect a wide range of lung reactions to injury [10] However, the temporal relationship to drug use and cessation is a potential clue to causation, although, in the case of methotrexate (Fig 18), drug reactions may occur years after starting treatment. Differentiating infection from acute drug reactions, or acute interstitial pneumonia, may also be problematic and bronchoalveolar lavage is usually required in cases with radiological features of diffuse alveolar damage to exclude infection [11].

Images for this section:
**Fig. 1:** 68 year old male with rheumatoid related chronic pleural thickening and a small effusion: CXR showing blunting of the costophrenic recesses.
Fig. 2: CT of same patient as fig. 1 showing smooth diffuse pleural thickening adjacent to a small effusion.
Fig. 3: Same patient as fig. 1. T1 weighted axial MRIs showing benign/inflammatory pleural thickening which is low signal on both sequences and the small effusion.
**Fig. 4:** Same patient as fig. 1. T2 weighted axial MRI showing benign/inflammatory pleural thickening which is low signal on both sequences and the small effusion.
Fig. 5: CT with bilateral trapped lung secondary to marked visceral parietal pleural thickening with bilateral pneumothoraces and associated effusions in a 74 year old male with severe rheumatoid disease.
**Fig. 6:** CXR of a 45 year old female patient showing bilateral rheumatoid nodules of variable size. The appearances are indistinguishable from pulmonary metastases. Note left shoulder replacement for rheumatoid arthritis.
Fig. 7: CT from the same patient as fig. 6 showing multiple well defined nodules which are predominantly peripheral in location.
Fig. 9: RA-ILD with a UIP pattern. HRCT showing coarse fibrosis with subpleural reticulation, microcystic honeycombing and subsequent architectural distortion with tractional airway dilatation.
Fig. 10: CXR with volume loss and basal reticulation typical of interstitial fibrosis.
**Fig. 11:** HRCT from the same patient as fig. 10 showing a fibrotic NSIP pattern with bilateral ground glass attenuation superimposed on reticulation with architectural distortion.
**Fig. 12:** HRCT showing typical features of organising pneumonia with bilateral bronchocentric and subpleural consolidation with additional perilobular signs in a 42 year old female patient.
Fig. 13: HRCT showing widespread ground glass attenuation, diffuse centrilobular nodules and two clearly defined thin-walled cysts in the lower lobes typical of LIP.
Fig. 14: HRCT in a 42 year old male patient with rheumatoid and airway disease. Nontapering, dilated bronchi and bronchioles, in keeping with cylindrical bronchiectasis.
Fig. 15: CXR showing air trapping and hyperinflation in a 46 year old female patient with rheumatoid disease.
**Fig. 16:** HRCT from same patient as fig. 15 showing a mosaic perfusion pattern during inspiration as a result of hypoxic vasoconstriction due to obliterative bronchiolitis.
Fig. 17: Section from a CT of a 42 year old male with rheumatoid disease showing nodules in a perilymphatic and centrilobular distribution consistent with follicular bronchiolitis, confirmed by histology.
Fig. 18: HRCT with widespread ground glass opacity centrilobular nodules typical of subacute hypersensitivity pneumonitis, in this case provoked by initiation of methotrexate 4 months earlier. Subsequent imaging, following cessation of therapy, showed complete resolution.
Fig. 8: CT of bilateral cavitating rheumatoid nodules in a 36 year old male.
Conclusion

The thoracic manifestations of rheumatoid arthritis are common and potentially fatal. It is important that the radiologist is aware of potential diagnoses in order to guide early therapy.

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References


