Cryptogenic organizing pneumonia: Typical and atypical imaging features on high resolution computed tomography

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Authors: C. Dornia¹, D. Manos², J. L. Babar³, S. Feuerbach¹, O. W. Hamer¹, ¹Regensburg/DE, ²Halifax, NS/CA, ³Cambridge/UK
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Learning objectives

1. Learn the clinical background and pathophysiologic basis of Cryptogenic Organizing Pneumonia (COP)
2. Learn the classical HRCT appearance of COP
3. Learn how to recognize unusual patterns and avoid diagnostic pitfalls
4. Learn how to work up difficult cases

Background

Organizing pneumonia (OP) presents with variable imaging findings, but shows a typical histopathologic pattern. Although the pathologic features were identified over 150 years ago, it was not until 1983 that OP was established as a novel, independent entity (1, 2). Because the etiology was unknown the term "Cryptogenic Organizing Pneumonia" (COP) was chosen. However, in addition to the cryptogenic form numerous triggers have now been identified (3, 4 and 5), including infections, inflammatory bowel disease, drugs, collagen vascular disease, organ transplantation and radiation directed to the chest. The correct terminology recommended by the American Thoracic and European Respiratory Society is as follows: If a cause is evident the disease should be termed "Secondary Organizing Pneumonia", if the cause remains unknown the term "Cryptogenic Organizing Pneumonia" should be used. Cryptogenic and secondary OP cannot be distinguished on the basis of imaging. To avoid confusion with small airway disease, the term bronchiolitis obliterans organizing pneumonia (BOOP) as a competing name for OP has been discarded by the American Thoracic and European Respiratory Society in 2002 (6).

The mechanisms of disease and the pathogenesis are not entirely understood, but OP is considered to be a nonspecific repair process of the lung in response to various kinds of lung insults. The histopathologic hallmark of OP is intra-alveolar granulation tissue plugs (Masson bodies) which extend into the bronchiolar lumen. The presence of these plugs is a prerequisite for restoration of tissue integrity (7).

Clinically OP is characterized by an indolent onset of a flu-like illness including cough, fever, and dyspnoea. Less common symptoms are night sweats, weight loss and haemoptysis. In the vast majority OP shows an excellent response to corticosteroid treatment. 70-80% of patients show rapid clinical improvement and resolution of radiological findings under corticosteroid treatment. However, long term treatment is recommended due to the high risk of relapse if treatment lasts less than one year (7).
Imaging findings OR Procedure details

In typical cases, OP can be suspected on chest radiogram when bilateral patchy areas of consolidation with a tendency to progress and change location over time are seen. However, high resolution computed tomography (HRCT) of the chest is the modality of choice for the diagnosis of OP due to its higher sensitivity and specificity.

Classical Imaging Findings

The classical HRCT features of OP are consolidations (which are present in about 90% of cases) in a predominantly peripheral or peribronchovascular distribution (6). Air bronchograms are common (1). Ground glass opacities are present in about 60% of cases, usually associated with consolidation. Pleural effusions are rare. The lower zones of the lung seem to be involved more frequently (6). Figure 1 on page 11 and Figure 2 on page 14.

Less Common and Atypical Imaging Findings

Focal Lesions

Approximately 15% of patients with OP present with focal, mass-like consolidations or large nodules (> 1 cm) which can be solitary or multiple. These nodules are usually smoothly marginated, but spiculae and pleural tags are found in 35% and 38%, respectively (6). Exclusion of malignancy cannot be made on the basis of the imaging appearance alone. In most cases biopsy or resection is necessary (3). Figure 3 on page 13.

Crazy Paving

The so-called crazy paving pattern appears as thickened interlobular septa and intralobular lines superimposed on a background of ground-glass opacity (8, 9 and 10). The pattern was first described for alveolar proteinosis but has a wide differential diagnosis including OP. Figure 4 on page 12.

Linear and Band-like Morphology

A linear or band-like pattern of OP may occur in isolation or in combination with other patterns. On HRCT lines or bands longer than 2 cm are seen (3). Two distinct types have been described. The type-1 opacity extends in a radial manner along the line of the bronchi towards the pleura; the type-2 opacity occurs in a peripheral location, runs parallel to the pleura and bears no relationship to the bronchi. Most lesions ultimately contact the pleura (11). Figure 5 on page 15.

Perilobular Pattern
The perilobular pattern consists of opacities distributed around the secondary lobule with an arcadelike or polygonal appearance (3). This pattern is indicative of OP but not specific for OP (12) and is seen in up to 57% of cases (13). Figure 6 on page 16.

Reversed Halo Sign

The reversed halo sign appears as central ground glass opacity surrounded by a more dense, crescentic or ring shaped consolidation. Kim et al. found this pattern in 19% of patients with biopsy-proven OP (14). However, it is not specific for OP as it has been described in cases of pulmonary infections, Wegener’s granulomatosis and lymphomatoid granulomatosis (15, 16 and 17). The term derives from the halo sign, in which ground glass opacity extends around the circumference of a central nodule or mass as seen for instance in angioinvasive aspergillosis. Figure 7 on page 17.

Differential Diagnosis

There are several differential diagnoses to consider when OP is suspected. If the HRCT findings are dominated by consolidation and ground glass opacity, pneumonia, bronchioloalveolar carcinoma and pulmonary infarction are the most important differential diagnoses. If there are focal nodules and masses, bronchial carcinoma, metastases, sarcoidosis, Wegener’s granulomatosis and hypersensitivity pneumonitis have to be considered (1, 6).
Images linked within the text of this section:

**Fig.**: HRCT image of the lung shows bilateral areas of consolidation with peripheral distribution (arrows). Positive air bronchograms are clearly visible. The morphology and the distribution are typical for OP, which was proven by biopsy.
**Fig.**: HRCT image of the lung shows areas of consolidation in both upper lobes (straight arrows). In addition ground glass attenuation is seen on the fringe of the consolidations (curved arrows).
Fig.: HRCT. 4 cm sized nodule with spiculae and pleural tag in the left lower lobe in a male patient with OP (curved arrow). Additionally small focal consolidation in the anterior basal segments of both lower lobes and ground glass opacities in the middle lobe (straight arrows) are seen.
Fig.: HRCT. Consolidation and ground glass opacity are seen in both upper lobes and the superior segment of the right lower lobe. Crazy paving pattern is noted in the right upper lobe (arrow).
**Fig.:** HRCT. Band-like consolidation in the right upper lobe (curved arrow). The lesion runs parallel to the pleura and bears no relationship to the bronchi (type-2 opacity). Additionally focal ground glass attenuation is seen in both upper lobes (straight arrows).
**Fig.**: HRCT of the lung in a male patient with ulcerative colitis on sulfasalazine and secondary OP. Bilateral foci of consolidation with peripheral (curved arrows) and perilobular (straight arrows) distribution. Image courtesy of NL Müller, Department of Radiology, Vancouver General Hospital, University of British Columbia, Canada.
Fig.: HRCT. Reversed halo sign with central ground glass opacity (asterisk) and surrounding ring shaped consolidation (arrows) in the left lower lobe. The sign is characteristic but not specific for OP.

Additional images for this section:
Fig. 1: HRCT image of the lung shows bilateral areas of consolidation with peripheral distribution (arrows). Positive air bronchograms are clearly visible. The morphology and the distribution are typical for OP, which was proven by biopsy.
Fig. 2: HRCT. Consolidation and ground glass opacity are seen in both upper lobes and the superior segment of the right lower lobe. Crazy paving pattern is noted in the right upper lobe (arrow).
Fig. 3: HRCT. 4 cm sized nodule with spiculae and pleural tag in the left lower lobe in a male patient with OP (curved arrow). Additionally small focal consolidation in the anterior basal segments of both lower lobes and ground glass opacities in the middle lobe (straight arrows) are seen.
**Fig. 4:** HRCT image of the lung shows areas of consolidation in both upper lobes (straight arrows). In addition ground glass attenuation is seen on the fringe of the consolidations (curved arrows).
**Fig. 5:** HRCT. Band-like consolidation in the right upper lobe (curved arrow). The lesion runs parallel to the pleura and bears no relationship to the bronchi (type-2 opacity). Additionally focal ground glass attenuation is seen in both upper lobes (straight arrows).
**Fig. 6:** HRCT of the lung in a male patient with ulcerative colitis on sulfasalazine and secondary OP. Bilateral foci of consolidation with peripheral (curved arrows) and perilobular (straight arrows) distribution. Image courtesy of NL Müller, Department of Radiology, Vancouver General Hospital, University of British Columbia, Canada.
Fig. 7: HRCT. Reversed halo sign with central ground glass opacity (asterisk) and surrounding ring shaped consolidation (arrows) in the left lower lobe. The sign is characteristic but not specific for OP.
Conclusion

The classical HRCT features of OP are well known. However, numerous atypical patterns have also been described. A thorough understanding of the various appearances of OP is a prerequisite for image interpretation.

Personal Information

Christian Dornia, MD
Department of Radiology
University Hospital
93053 Regensburg, Germany
christian.dornia@klinik.uni-regensburg.de

References


