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PULMONARY SARCOIDOSIS

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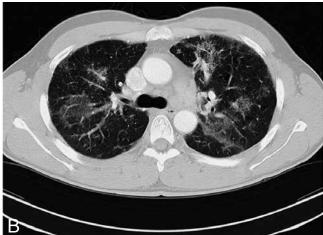
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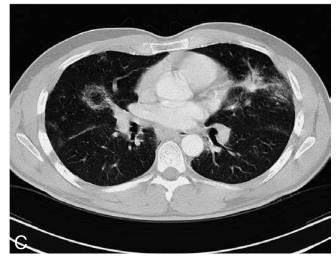
Background: A 34-year-old male teacher was referred to the hospital with a persisting dry cough and dyspnea on exercise since eight weeks. He had no fever, neither complaints of ear, nose or throat. There were no complaints during the night. He had been a smoker until four months before presentation (12 pack years). At work a student was diagnosed with pulmonary tuberculosis, but the Mantoux and Quantiferon tests were negative. Physical examination was normal, without fever, lymphadenopathy or auscultation abnormalities.

Laboratory investigation revealed a C-reactive protein of 2 mg/L. Pulmonary function testing showed a slight restriction. Immunological bronchial alveolar lavage (BAL) was rich of cells, especially T-lymphocytes of the CD4 type. CD4+/CD8+ ratio of the BAL was raised to 4.2, compared to a ratio of 2.4 in blood. There were no eosinophils found in the BAL.

Conventional chest radiographs were performed, and showed multiple areas of consolidation in the bilateral lung fields, predominantly on the right side.







Work-up

HRCT scan of the thorax (pulmonary window setting) (Fig. 1) shows multiple areas of groundglass opacities in both lungs, some of them combined with surrounding crescent and ring shaped consolidation, representing the 'reversed halo sign'.

Radiological diagnosis

The diagnosis of *pulmonary sarcoidosis* was based on clinical history, the bronchial alveolar lavage and the radiological finding of the reversed halo sign on HRCT-thorax.

Discussion

The reversed halo sign is characterized by the existence of central ground-glass opacities surrounded by crescent or ring-shaped consolidation.

Important clues in history and additional investigation are needed to make a final diagnosis. In this case the young age of the patient and the ongoing complaints without raised infection parameters were very helpful. The immunological bronchial alveolar lavage with a high CD4+/CD8+ ratio count was very suggestive for the diagnosis sarcoidosis. The presence of non-caseating granulomas in transbronchial biopsies was probative. In this case biopsies were taken from the left lower lobe, which showed granulomatous inflammation without necrosis. Auramin and Ziehl Neelsen stain were negative.

The reversed halo sign was first described in 2003 and associated with the diagnosis of cryptogenic organizing pneumonia (COP). An identical sign was earlier described as the atoll sign, and associated with bronchiolitis obliterans with organizing pneumonia (BOOP).

A similar sign, the 'fairy ring sign' was associated with sarcoidosis in 1999, but consisted in annular shadows around a normally aerated area in bilateral lungs and is thus different from the reversed halo. Although previously considered specific for

the diagnosis COP, in the past few years the reversed halo sign was linked to several other diagnoses and a connection with sarcoidosis is emphasized. The mechanism of development of the RHS in sarcoidosis is still unclear.

Besides sarcoidosis, BOOP and COP, other diagnoses that should be considered if a reversed halo is seen are organizing pneumonia, pneumocystis jiroveci pneumonia (PJP), invasive pulmonary aspergillosis, tuberculosis, Wegener granulomatosis, histoplasmosis, cryptococcosis, pulmonary embolism, edema, lepidic predominant adenocarcinoma (formerly bronchioalveolar carcinoma), zygomycosis, pulmonary paracoccidioidomycosis, and lipoid pneumonia or lymfomatoid granulomatosis.

The presence of nodular walls or nodules inside the reversed halo is highly suggestive of granulomatous diseases. Other radiological signs on HRCT that could support the diagnosis pulmonary sarcoidosis are intrathoracic lymphadenopathy (most common) hilar and right paratracheal, the presence of small nodules in a perilymphatic distribution, multifocal parenchymal opacities (10-20%) including the sarcoid galaxy sign and extensive fibrosis (20-25%) particularly in the parahilar region.

Bibliography

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