LYMPHOMATOID GRANULOMATOSIS

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Key-word: Lymphomatoid granulomatosis

Background: A 70-year-old non smoker man presented to the pneumologist with persistent dyspnea.

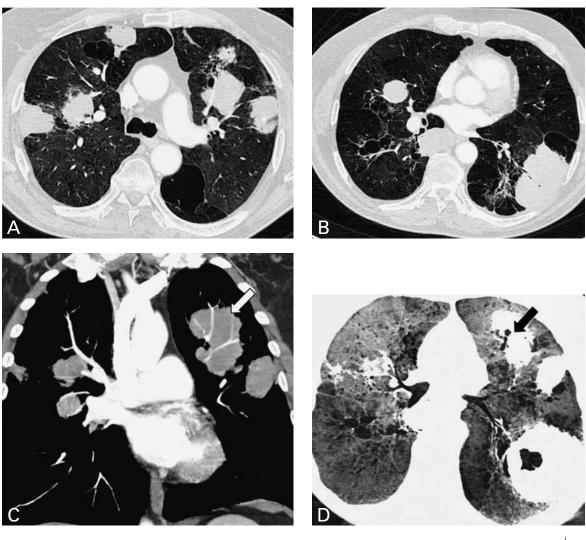


Fig. $\frac{1A}{1C} \frac{1B}{1D}$

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Work-up

CT scan of the thorax (Fig. 1) shows on axial slices (lung window setting) (A,B) multiple well-defined nodules and masses in all lung zones. Maximum intensity projection (MIP) reformatted image in the coronal plane (C) shows preserved pulmonary vessels inside a lesion in the left upper lobe (arrow). Minimum intensity projection (MIP) axial view (D) demonstrates dilated bronchus in a lesion located in the lingula (arrow) and cavitation of a lesion in the left lower lobe.

Radiological diagnosis

Following transthoracic biopsy under CT guidance, the diagnosis of *lymphomatoid granulomatosis* was achieved.

Discussion

Lymphomatoid granulomatosis (LG) was described for the first time in 1972 by Liebow. LG represents 3% of the primary pulmonary lymphomas which represent 3-4% of the extranodal non Hodgkin lymphomas.

LG is a polymorphic lymphoid infiltration invading the vascular structures and leading to angiocentric necrosis of the lung parenchyma. The infiltration is composed by large atypical CD20+ B-cells showing the LMP protein of the Epstein Barr Virus, dispersed together with small reactive T-cells.

There are three subcategories of LG based on the proportion of the transformed B-cells. Grade 1 shows the lowest and grade 3 the highest amount of atypia. Grades 2 and 3 are considered to be malignant.

The symptoms are mainly respiratory, including dyspnea, cough, thoracic pain and hemoptysis. Fever and weight loss can also be present. Beyond lungs, central and peripheric nervous systems, skin, kidneys and mediastinal lymph nodes can be involved. Men and women are equally reached at a

mean age of 55 years. On chest radiography the lesions appear like nodular opacities with clear or blurred outlines, commonly predominating in the lower lobes. They have a tendency to confluence and excavate, and sometimes to migrate or disappear. On chest CT LG presents as nodular lesions localized along bronchovascular structures, that are multiple in 75% and bilateral in 60 to 80%. Most nodules measure less than 1 cm but diameters up to 10 cm have been reported. The lesions may show preserved air bronchogram and vessels inside the lesions. Distended bronchi inside the lesions have been suggested to be evocative of the diagnosis but the mechanism is still unexplained. Central necrosis is possible with formation of thin-walled cystic lesions. Hilar or mediastinal lymph nodes can be present.

The differential diagnosis of multiple pulmonary nodules occasionally presenting with excavation includes: abscesses, septic emboli, Wegener or other granulomatosis, excavating pulmonary neoplastic lesions, necrotising sarcoid granulomatosis, and less frequently BALT lymphoma, Churg-Strauss disease and organizing pneumonia.

The diagnosis is not based on imaging but on pathology including demonstration of the Epstein Barr Virus in situ hybridation.

Bibliography

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