Cystic disease with sparing of lung bases

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A 39-year-old male smoker presented to the outpatient clinic with complaints of dry cough and dyspnea on moderate exertion. Laboratory test results were unremarkable. Chest CT showed multiple bilateral pulmonary cysts, predominantly in the upper lung fields (Figure 1).

Pulmonary cysts are characterized on CT as rounded, low-attenuation areas in the lung parenchyma, with a well-defined interface with the adjacent normal lung. The cyst wall can vary in thickness but is typically thin. The cysts usually contain air but can sometimes contain fluid. A cystic pattern is seen in a number of diseases, the most characteristic of which are lymphangioleiomyomatosis, Langerhans cell histiocytosis (LCH), lymphocytic interstitial pneumonia, and Birt-Hogg-Dubé syndrome (BHDS).

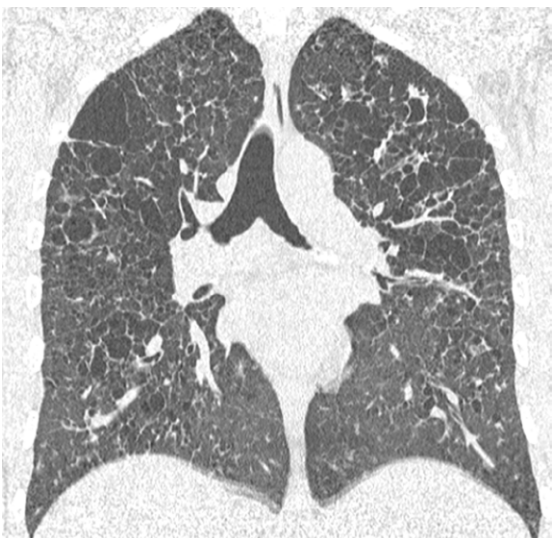

Figure 1. Coronal CT reconstruction showing multiple irregularly shaped cysts, predominating in the upper lung fields. Note the relative sparing of the lung bases.

Some CT criteria can be used in differential diagnosis. In lymphocytic interstitial pneumonia, cysts are less numerous and can be accompanied by ground-glass opacities. Two syndromic conditions can present with pulmonary cysts and renal masses: tuberous sclerosis and BHDS. In BHDS, the cysts are less numerous and larger and predominate in the lower lobes. In tuberous sclerosis, the cysts represent lymphangioleiomyomatosis, are more numerous and diffuse, and also affect the lung bases. In LCH, the cysts can be bizarrely shaped and, most importantly, they predominate in the upper lung fields, with sparing of the lung bases, especially the costophrenic sulci.

LCH is a rare disorder of unknown origin, characterized by an abnormal non-malignant proliferation of monoclonal Langerhans cells (histiocytes).It remains controversial whether LCH is a neoplastic or an inflammatory disorder. Pulmonary LCH is seen almost exclusively in cigarette smokers. Clinically, patients can be asymptomatic or present with cough and dyspnea. In many cases, the disorder is discovered either incidentally on routine examinations or because of complications, such as pneumothorax.[1,2]

In early stages of LCH, CT shows centrilobular nodules, which correspond to granulomas. They tend to cavitate, progressing to cysts. The cysts can be irregularly and bizarrely shaped, which differentiates them from the regular cysts in lymphangioleiomyomatosis. The lesions predominate in the upper lobes and spare the lung bases. In end-stage LCH, there may be only diffuse large irregular cysts without nodules.[1,2]

The finding of irregularly shaped cysts, predominantly in the upper lung fields, with sparing of the costophrenic sulci, and accompanied by small nodules, is highly suggestive of a diagnosis of LCH, generally without the need for lung biopsy, which is reserved for atypical cases. In the case of our patient, the final diagnosis was LCH.

REFERENCES

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