Unusual Findings of Langerhans Cell Histocytosis in a Young Asymptomatic Patient: Case Report

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Abstract

Cyst is described on high-resolution computed tomography (HRCT) as focal round area of parenchymal hyperlucency with a well-defined interface with normal lung. A relatively small subset of lung diseases should be suspected when cystic pattern is seen on HRCT. In particular, the differential diagnosis of this pattern relies upon features such as: cranio-caudal distribution, cystic shape, and coexisting pulmonary or extra-pulmonary findings.

Pulmonary Langerhans Cell Histocytosis (PLCH) is a smoke-related disease associated with cystic pattern. PLCH is characterized by infiltration of Langerhans cells in pulmonary interstitium with varying pathologic findings and depend on disease stage. The findings usually consist of a heterogeneous combination of lung nodules and cysts with upper lobes predominance. Notably, the HRCT pattern of PLCH is often striking and usually suggests confident diagnosis. Nevertheless, cases with atypical features can be seen.

This case report describes the ambiguous imaging findings of a biopsy-proven PLCH.

Case Report

A 33 years-old smoker male presented at the emergency department with right abdominal pain. A heterogeneous left renal mass was reported at ultrasonographic examination of the abdomen. The left incidental finding was therefore investigated by both Magnetic Resonance Imaging (MRI) and CT of the abdomen (Figure 1). The MRI and CT highlighted fatty areas within the left renal mass. Besides, the upper CT images revealed the presence of bilateral basal pulmonary cysts.

Figure 1A-B: Incidental left renal mass. A, axial MRI T2 image showing iperintense area (arrow) in the upper portion of left kidney. B, axial post-contrast CT image showing low density area (arrow) in the upper portion of left kidney, with central contrast enhancement (asterisk).

Figure 2A-D: A-B, axial CT image at the level of costo-phrenic sulci. Bilateral cysts are seen (arrows) involving costo-phrenic sulci down to their deeper portion. The cysts present homogeneous size and shape. C-D, sagittal (C) and coronal (D) CT image allow panoramic view of pulmonary cysts distribution. Involvement of lower lobes including costo-phrenic sulci (arrows) is shown in association with relative sparing of upper lobes.

Therefore an HRCT scanning of the whole lung was added and the presence of multiple thin-walled cystic lesions with irregular shape was then confirmed (Figure 2) [1-3]. The cysts were homogeneous in size

and shape. Notably, such pulmonary cysts showed a slightly predominant distribution in the lower lobes with conspicuous involvement of the costophrenic sulci. No pulmonary nodules were identified in the lung parenchyma [4-6].

Bronchoalveolar Lavage (BAL) and transbronchial biopsy were performed but were not diagnostic. Therefore, a surgical lung biopsy was performed. The histologic assessment of the sample showed Langerhans cells infiltration of the pulmonary interstitium.

In addition, the patient underwent ultrasonography-guided fine needle biopsy of the renal mass, which showed neoplastic cell and adipocytes. The renal mass turned out to be a renal clear cell carcinoma that was therefore resected by laparoscopic tumorectomy.

Discussion

HRCT features of PLCH consist of a combination of centrilobular ill-defined nodules (2-10 mm in diameter) and cysts of variable shape and size (usually <10 mm) [7,8]. In advanced stages of PLCH, cysts may enlarge or coalesce resulting into bizarre-shaped cystic lesions [7]. All of these abnormalities mainly involve the upper and the middle lung zone, sparing the costo-phrenic sulci. Also, the cranio-caudal distribution of cysts is usually helpful in the differential diagnosis of cystic lung diseases.

This report shows a case of PLCH with unusual distribution of cysts within lung parenchyma and coexisting fatty renal lesion. Indeed, lung cysts were extraordinarily predominant in the lower lung zone, with conspicuous involvement of costo-phrenic sulci. Such a distribution pattern is atypical for PLCH, whereas it is typical in other cystic lung diseases such as Lymphangioleiomyomatosis (LAM) or Birt-Hogg-Dubé syndrome [7,9,10]. Notably, involvement of costo-phrenic sulci has been reported in children with pulmonary PLCH [8,11]. Hence, we might hypothesize infantile onset of asymptomatic PLCH in the reported case. Furthermore, this individual showed homogeneous shape and size of the lung cysts that, again, are unusual in PLCH. However, in this case, PLCH was first suspected by the radiologist because of the patient's gender and smoking history.

Cases of cystic lung diseases with overlapping features have been described in the literature. Keyzer et al. [12] described a case of LAM presenting with cough and mild dyspnea in a 26 years-old heavy smoker female. As opposite to ours, HRCT findings were suggestive of PLCH: irregular cysts and nodules (both solid and cavitated) with a predominant apical distribution.

The renal mass characterized by fatty areas could have been diagnosed as angiomyolipoma on MRI and CT. Notably, angiomyolipoma is a common abdominal finding in LAM. Therefore, the homogeneous distribution of cysts seen in this subjects and the presence of fatty benign renal lesion would be consistent with LAM, especially in young female [13-15]. Nevertheless, renal neoplasms with small fat spots have also been described in Birt-Hogg-Dubé Syndrome [16].

In conclusion, this case report shows that unusual distribution of cysts may be observed in PLCH. Moreover, abdominal findings might overlap ancillary side findings of cystic lung diseases other than PLCH. Therefore, lung biopsy is warranted to secure reliable diagnosis of cystic lung disease in individuals with contradictory radiological findings [7,17,18].

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