Case Report

Asymptomatic lymphangioleiomyomatosis: Large cyst mimicking a hydropneumothorax in a healthy patient

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ABSTRACT

Lymphangioleiomyomatosis (LAM) is a rare, idiopathic, cystic disease that affects lungs of young women at childbearing age. Usually, LAM clinical manifestations are pneumothorax, progressive dyspnea and chylous pleural effusions. In many cases, due to unusual and non-specific symptoms, LAM is mis-recognized and patients, who are affected by such disease, receive delayed diagnosis. This case report focuses on a 45-year-old woman patient with asymptomatic lymphangioleiomyomatosis who presented a big cyst which makes it looks like hydropneumothorax condition. Although chest radiograph and following CT scan has given an incidental diagnosis of right hydropneumothorax, treatment with drainages and video-assisted thorascopic surgery, instead, has allowed us to formulate correct diagnosis of cysts in LAM.

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Introduction

Lymphangioleiomyomatosis (LAM) is uncommon interstitial lung disease affecting young women during their reproductive years. It is microscopically characterized by 2 phenomena: first, abnormal proliferation of smooth muscle-like cells which determines progressive obstruction of small airways and cystic changes; second, extrapulmonary manifestations such as lymphangiomomas and renal angiomyolipomas [1-2].

In addition, LAM is more likely to be present in patients with tuberous sclerosis complex (TSC) which is an autosomal dominant syndrome characterized by hamartoma formation within multiple organs, cerebral calcifications, seizures and cognitive defects [3-4].
Generally, patients with LAM present progressive dyspnea, chylous collection, recurrent pneumothorax and occasionally hemoptysis [5]. Recommended imaging technique is chest computed tomography (CT) scan which is able to detect the presence of well circumscribed, rounded and thin-walled scattered cysts in bilateral roughly symmetric pattern without any lobar predominance [6].

Case report

A 45-year-old woman patient with history of multinodular goiter come to the endocrinology department of our hospital. The patient presented stable vital parameters as tested by successive blood examinations, certifying good health condition overall. Although, the patient had no symptoms of fever, cough or dyspnea, when she underwent the pre-operative chest radiograph for thyroidectomy, an incidental right-sided hydropneumothorax was found (Fig. 1).

An ultrasonography examination revealed presence of particulate matter inside the pleural effusion at the base of the right lung. Then, the patient underwent another examination made with the aid of chest CT scan. Diagnostic analysis was made both under decubitus normal as well as prone position which evidenced a large right-sided loculated hydropneumothorax that did not change by varying the decubitus (Figs. 2A-B, 3).

A chest drain (24 CH) was placed on the sixth right intercostal space with the aid of thoracic surgeon for reducing the hydropneumothorax.

However, chest drainage failed due to resistance of hydropneumothorax even after needle aspiration was applied. Hence, we decided to adopt subscapular drainage (12CH UNICO) under ultrasonography guidance. Then, the purulent fluid was drained (500 mL of yellow-brown secretion).

Once that drainage was executed, a second chest CT scan was performed and it showed the persistence of right lung atelectasis on the base.

Furthermore, video-assisted thorascopic surgery was carried out and exhibited a big cystic lesion with a diameter of 18 × 12 cm in the middle lobe of the right lung (Fig. 4). Hence, this bulky cyst was removed and analyzed.
LAM is considered rare disease that can be fatal due to abnormal proliferation of smooth muscle cells in the lung parenchyma and widespread cystic destruction of lungs [7-8]. Common LAM symptoms include dyspnea, recurrent pneumothorax, chylous collections, and hemoptysis [2,5].

Typically, chest radiograph illustrates hyper inflated lung due to obstructive nature of LAM and, in later stages, reticular pattern from the coalescence of the cysts [9,10]. Highly recommended imaging technique (HRCT) exhibits multiple thin-walled cysts in both lung parenchyma with pleural effusion, pneumothorax, ground glass opacities and pericardial effusion [6,9,11].

In addition, HRCT represents first stage of investigation related LAM once suspicious arouse, as suggested by European Respiratory Society (ERS) and American Thoracic society (ATS) guidelines [12-13]. In contrast, lack of typical CT features excludes LAM diagnosis.

Usually, LAM diagnosis is upheld by lung biopsy with immunochemical strains (HMB-45) that are specific for smooth muscle cells [14–15].

As demonstrated so far, after many unsuccessful attempts to drain the hydropneumothorax, video-assisted thorascopic surgery revealed to be appropriate technique that attested the presence of the cyst on the right side of the lung and permitted its removal by surgery. Finally, valid histological research confirms the diagnosis of unusual LAM.

**Patient consent**

The authors obtained from the patient written informed consent for the publication of this case report and images.

**Authors’ contributions**

The conception and design of the study: CB and UB; Manuscript preparation and manuscript editing: RM and AM; Acquisition of data: CR and AM; Literature research and drafting the article: CA and RM; Revising it critically for important intellectual content: UB and CB; Responsible for the surgical intervention: FM; Final approval: All authors.

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