Case Report

Pulmonary mucosa-associated lymphoid tissue (MALT) lymphoma presented as cystic lung disease – a case report in Tuen Mun Hospital Hong Kong

P.Y.L. Chan, FRCR, MBBS*, A. Li, FHKCR, FCR

Department of Radiology and Nuclear Medicine, Tuen Mun Hospital, Hong Kong

A R T I C L E   I N F O

Article history:
Received 1 May 2022
Revised 18 June 2022
Accepted 21 June 2022

Keywords:
Pulmonary lymphoma
Pulmonary MALToma
Cystic lung disease

A B S T R A C T

We report a rare case of pulmonary mucosa-associated lymphoid tissue lymphoma (MALToma) in a 52-year-old woman presented as cystic lung disease together with ground-glass lesion on computed tomography (CT) of the thorax incidentally found as part of workup for organ donation.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Pulmonary mucosa-associated lymphoid tissue lymphoma (MALToma) is a rare, but the most common form of primary pulmonary lymphoma, accounting for up to 90% of cases [1]. Common presentations of pulmonary MALToma include consolidations and multiple bilateral pulmonary nodules [2]. Associated features such as air bronchogram and cystic bronchiectasis are also common. Some report reticular interstitial lung disease pattern [3]. Manifestation as diffuse cystic lung disease in pulmonary MALToma is rare. We herein report a case of a 52-year-old lady without significant past medical history who was diagnosed incidentally to have cystic lung disease with co-existing ground-glass lesion which was proven to be pulmonary MALToma on wedge resection of the lung lesions.

Case report

A 52-year-old woman presented to the respiratory team after CT volumetry of the liver, as part of workup for liver donor, revealed multiple thin-walled lung cysts of variable sizes in the lung bases. She was rejected as a liver donor due to suspected malignancy in the lungs. This occurred after the onset of the COVID-19 pandemic and she had received multiple doses of the COVID-19 vaccination throughout the period. There was no evidence that she ever contracted the virus. The patient

* Competing Interests: All authors declare no conflict of interest.
* Corresponding author.
E-mail address: pris0727@gmail.com (P.Y.L. Chan).
https://doi.org/10.1016/j.radcr.2022.06.064
1930-0433/© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)
was a never-smoker and never-drinker, had no significant past medical history of autoimmune-related disease or infection and no chronic illness such as diabetes mellitus and hypertension. She did not have raise inflammatory or tumor serum markers. Her complete blood profile with differential counts also did not reveal any significant abnormality.

Chest X-ray did not reveal any significant abnormality. A dedicated CT scan of the thorax with intravenous contrast was performed. It revealed multiple thin-walled lung cysts of variable sizes ranging from 4 mm to 25 mm in size. The cysts showed lower lobe predominance with involvement of the costophrenic sulci (Figs. 1A–C). In addition, there was a ground-glass lesion measuring 1.8 × 1 cm (axial transverse × antero-posterior dimension) seen at the inferior aspect of right upper lobe posterior segment abutting the right oblique fissure with mildly spiculated margin (Fig. 2). Another smaller ground-glass lesion was also seen in the medial basal segment of right lower lobe.

No consolidation or features of interstitial lung disease was demonstrated. Few tiny nonspecific nodules were seen scattered in the left lung. There was no associated thoracic lymphadenopathy or pleural effusion. Subsequent bronchoscopy did not reveal any endobronchial lesion and septic workup for the bronchial aspirate was also negative.

The concerned right upper lobe ground-glass lesion was then resected alongside one of the right lower lobe cyst via video-assisted thoracotomy to obtain histopathological diagnosis. The right upper lobe ground-glass lesion was localized using indocyanine green-lipiodol emulsion via hookwire prior to operation (Fig. 3). On histopathology, both of these lesions demonstrated atypical aggregates of lymphoid proliferation, with the final diagnosis being extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (Fig. 4).

The patient had been asymptomatic all along and had thus been referred to the hematology team for further workup. The latest CT scan of the thorax prior to treatment showed interval enlargement of the lung cysts, while the smaller ground-glass lesion seen in the medial basal segment of right lower lobe was similar. PET-CT performed showed only minimal FDG activity in the lesions and did not reveal any other focal hypermetabolic lesion to suggest nodal or extralymphatic lymphoproliferative disease. The patient is currently on conservative treatment as she is asymptomatic and no other organs are involved.

**Discussion**

Due to a lack of specific biological serum markers and nonspecific wide range of radiological features, pulmonary MAL-Toma often has a delayed diagnosis. Patients are also usually asymptomatic or present with nonspecific clinical signs and symptoms. Wen et al [2] reported consolidation as the most frequent CT feature. All of the patients with consolidations had air bronchogram, while most of them had mild bronchiectasis. Ground-glass nodules and pulmonary cysts accounted for only 6% of the cases reported in the study [2].

Bae et al. also reported single nodular or consolidative pattern as the most common manifestation of pulmonary MAL-

---

*Fig. 1 – (A–C) Axial CT of the thorax of this 52-year-old lady showing thin-walled cysts of variable sizes (white arrows) distributed in both lungs with lower lobe predominance involving the lung base.*
Toma. In addition, diffuse interstitial lung disease was also reported as one of the possible radiological features [3].

Other diseases that present as diffuse cystic lung disease such as lymphangioleiomyomatosis and Langerhans cell histiocytosis were also considered in our case but the radiological pattern was not typical of those diseases. Our patient also had no history of HIV infection, autoimmune related disease and had been asymptomatic all along. It proved to be a diagnostic challenge and highlighted the importance to consider alternative diagnosis such as pulmonary MALToma in cases of cystic lung disease that do not fit into the typical radiological pattern of lymphangioleiomyomatosis or Langerhans cell histiocytosis.

Lymphocytic interstitial pneumonia is another rare differential diagnosis of diffuse cystic lung disease that could also exhibit abnormal lymphoid aggregate proliferation. Associated diseases such as Sjogren syndrome, systemic lupus erythematosus and HIV infection are usually present [4]. Lymphocytic interstitial pneumonia and lymphoma belong to a spectrum of pulmonary lymphoproliferative disease that also includes diseases such as follicular bronchiolitis and focal lymphoid hyperplasia [5]. Cystic lung disease when associated with ground-glass nodules demonstrating perilymphatic distribution should lead radiologists to raise concern of underlying pulmonary lymphoproliferative disease.

Conclusion

We hope that this case illustrates the importance of considering the rare differential diagnosis of pulmonary MALToma as an atypical differential diagnosis of cystic lung disease on CT imaging to ensure timely diagnosis and management in a multidisciplinary approach.

Patient consent

An informed consent for the case report titled “Pulmonary mucosa-associated lymphoid tissue (MALT) lymphoma presented as cystic lung disease – a case report in Tuen Mun Hospital Hong Kong” has been taken on 19th April, 2022 with the patient herself. A signed informed consent form has been obtained and retained by the first author.

References

[1] Primary Pulmonary Lymphoid Lesions: Radiologic and Pathologic Findings | RadioGraphics [Internet]. [cited 2022]
Feb 7]. Available from: 
https://pubs.rsna.org/doi/10.1148/rg.2016140339.

[2] Deng W, Wan Y, Yu J. Pulmonary MALT lymphoma has variable features on CT. Sci Rep 2019;9(1):8657.

[3] Bae YA, Lee KS, Han J, Ko Y-H, Kim B-T, Chung MJ, et al. Marginal zone B-cell lymphoma of bronchus-associated lymphoid tissue: imaging findings in 21 patients. CHEST 2008;133(2):433–40.

[4] Cha S-I, Fessler MB, Cool CD, Schwarz MI, Brown KK. Lymphoid interstitial pneumonia: clinical features, associations and prognosis. Eur Respir J 2006;28(2):364–9.

[5] Baldi BG, Carvalho CRR, Dias OM, Marchiori E, Hochhegger B. Diffuse cystic lung diseases: differential diagnosis. J Bras Pneumol Publ 2017;43(2):140–9.