Respiratory Medicine Case Reports 28 (2019) 100912

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Smoking-related diffuse cystic lung disease

Ali Ataya[a,*], Leonard Riley[a], Kristianna Fredenburg[b], Mark Brantly[a]

[a] University of Florida, Division of Pulmonary, Critical Care and Sleep Medicine, Gainesville, FL, USA
[b] University of Florida, Department of Pathology, Gainesville, FL, USA

A B S T R A C T

Smoking tobacco is associated with an array of pulmonary symptoms and diseases. We describe a case of a woman with a spontaneous pneumothorax and diffuse cystic lung disease due to smoking. The presence of diffuse cystic changes in a woman is suggestive of lymphangioleiomyomatosis (LAM); however, her vascular endothelial growth factor-D was normal and surgical lung biopsy and pathology had notable absence of LAM cells and presence of intra-alveolar pigment laden macrophages and intraluminal mucostasis. Smoking-related diffuse cystic lung disease can mimic LAM and is a novel entity with only four other cases reported.

1. Introduction

Cysts are commonly seen on computed tomography of the chest and present a diagnostic challenge. The combination of pertinent positive and negative findings by clinical, radiographic, and pathologic evaluation help differentiate among these diseases. We present a case of diffuse cystic lung disease (DCLD) due to smoking.

2. Case report

A 46-year-old woman with an active, 25-pack-year history of tobacco use presented for acute onset of shortness of breath and right-sided pleuritic chest pain of one week's duration. She had no pertinent medical history or family history of pulmonary, neurologic, or renal disease. She has no history of recreational drug abuse. Physical exam revealed clear breath sounds diffusely and the absence of any cutaneous findings. Initial laboratory studies were unremarkable including complete blood count and complete metabolic panel. Chest radiograph demonstrated a right-sided pneumothorax with collapse of the right lower lobe. Computed tomography (CT) of the chest confirmed the right-sided pneumothorax and diffuse cystic lung disease without any other parenchymal or tracheobronchial findings (Fig. 1). Further laboratory investigation with vascular endothelial growth factor-D (VEGF-D) (279 pg/mL), alpha-1 antitrypsin level (223 mg/dL) with Pi*MS genotype, ANA, and anti-SS-A/SS-B were unremarkable. A CT of the abdomen also lacked the presence of any renal lesions. She then underwent a VATS-guided surgical lung biopsy of the right upper and lower lobes with mechanical pleurodesis. Pathology exam revealed intraparenchymal cystic changes, abundant intra-alveolar pigment laden macrophages, respiratory bronchiolitis, and intraluminal mucostasis (Fig. 2). Immunohistochemical staining for lymphangioleiomyomatosis (LAM) and Pulmonary Langerhans Cell Histiocytosis (PLCH) were negative. There was absence of malignant cells, germinal centers and lymphoid follicles. With the aforementioned work up, the diagnosis of smoking-related diffuse cystic lung disease was ultimately reached.

3. Discussion

Diffuse cystic lung disease (DCLD) among smokers usually occurs in the setting of PLCH. The presence of a spontaneous pneumothorax and DCLD without parenchymal findings in a woman of childbearing age is highly suggestive of LAM; however, the normal VEGF-D level, absence of renal lesions, and absence of LAM cells on pathology argued against this. This case highlights how imperative it is to not base the diagnosis of LAM solely on clinical and radiographic findings. Our CT findings in a young woman is concerning for LAM; however, in the absence any other confirmatory findings for LAM the diagnosis should not be made on CT alone [1,2].

Only four other patients with smoking-related DCLD have been reported [3]. The exact mechanism is unknown; however, bronchiolitis-induced airway narrowing resulting in check-valve-mediated distal airspace dilation and cyst formation and expression of enzymes leading to matrix remodeling have been postulated [4–6]. Treatment is unclear without formal investigative studies, but we followed a conservative approach with tobacco cessation, immunizations, education about avoidance of environmental hazards, and discussion of risks for developing recurrent pneumothoraces.

* Corresponding author. University of Florida, Division of Pulmonary, Critical Care and Sleep Medicine, 1600 SW Archer Rd, M452, PO Box 100225, Gainesville, FL, 32610, USA.
E-mail address: ali.ataya@medicine.ufl.edu (A. Ataya).

https://doi.org/10.1016/j.rmcr.2019.100912
Received 1 June 2019; Received in revised form 8 July 2019; Accepted 20 July 2019
Available online 22 July 2019

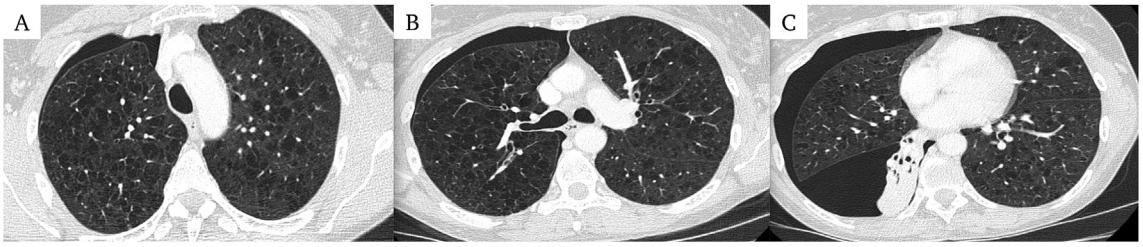

Fig. 1. Non-contrasted computed tomography of the chest in lung window and transverse plane demonstrating centrilobular emphysema and innumerable, diffuse cysts of variable sizes in upper (A), middle (B), and lower (C) lung zones.

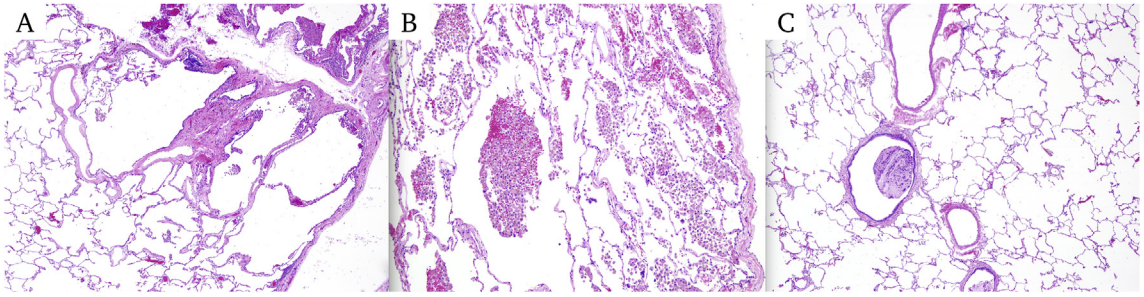

Fig. 2. Pathology showing intraparenchymal cystic changes (A), abundant intra-alveolar pigment laden macrophages (B), and intraluminal mucostasis (C).

Conflicts of interest

None.

Disclosures

None.

References

[1] F.X. McCormack, N. Gupta, G.R. Finlay, L.R. Young, A.M. Taveira-DaSilva, C.G. Glasgow, W.K. Steagall, S.R. Johnson, S.A. Sahn, J.H. Ryu, C. Strange, K. Seyama, E.J. Sullivan, R.M. Kotloff, G.P. Downey, J.T. Chapman, M.K. Han, J.M. D'Armiento, Y. Inoue, E.P. Henske, J.J. Bissler, T.V. Colby, B.W. Kinder, K.A. Wikenheiser-Brokamp, K.K. Brown, J.F. Cordier, C. Meyer, V. Cottin, J.L. Brozek, K. Smith, K.C. Wilson, J. Moss, Official American thoracic society/Japanese respiratory society clinical practice guidelines: Lymphangioleiomyomatosis diagnosis and management, 194 (6) (2016) 748–761.

[2] N. Gupta, G.A. Finlay, R.M. Kotloff, C. Strange, K.C. Wilson, L.R. Young, A.M. Taveira-DaSilva, S.R. Johnson, V. Cottin, S.A. Sahn, J.H. Ryu, K. Seyama, Y. Inoue, G.P. Downey, M.K. Han, T.V. Colby, K.A. Wikenheiser-Brokamp, C.A. Meyer, K. Smith, J. Moss, F.X. McCormack, Lymphangioleiomyomatosis diagnosis and management: high-resolution chest computed tomography, transbronchial lung biopsy, and pleural disease management. An official American thoracic society/Japanese respiratory society clinical practice guideline, Am. J. Respir. Crit. Care Med. 196 (10) (2017) 1337–1348.

[3] N. Gupta, T.V. Colby, C.A. Meyer, F.X. McCormack, K.A. Wikenheiser-Brokamp, Smoking-related diffuse cystic lung disease, Chest 154 (2) (2018) e31–e35.

[4] C. Rowan, D.M. Hansell, E. Renzoni, T.M. Maher, A.U. Wells, M.I. Polkey, P.K. Rehal, W.H. Ibrahim, G.N.M. Kwong, T.V. Colby, M. Pistolesi, F. Bigazzi, C.E. Comin, A.G. Nicholson, Diffuse cystic lung disease of unexplained cause with coexistent small airway disease: a possible causal relationship? Am. J. Surg. Pathol. 36 (2) (2012) 228–234.

[5] L. Kawano-Dourado, B.G. Baldi, O.M. Dias, F.D.C. Bernardi, C.R.R. Carvalho, M. Dolhnikoff, R.A. Kairalla, Scattered lung cysts as the main radiographic finding of constrictive bronchiolitis, Am. J. Respir. Crit. Care Med. 186 (3) (2012) 294–295.

[6] N. Gupta, R. Vassallo, K.A. Wikenheiser-Brokamp, F.X. McCormack, Diffuse cystic lung disease. Part II, Am. J. Respir. Crit. Care Med. 192 (1) (2015) 17–29.