中文題目：一位47歲女性的口香糖樣痰液

英文題目：Gum-like Sputum in a 47 Years Old Woman

作　　者：莊明哲[1],鄭文建[2]

服務單位：[1]中國醫藥大學附設醫院內科部,[2]中國醫藥大學胸腔內科

Introduction

Chyloptysis is a rare symptom and the differential diagnosis of chyloptysis includes lymphangioleiomyomatosis, yellow nail syndrome, or thoracic lymphangiectasis. We presented a case of a woman developed chyloptysis, improving after the sirolimus control.

Case report

This 47-year-old woman presented shortness of breath for one year and chylothorax (Triglyceride 4200 mg/dL in pleural effusion) was diagnosed 9 years ago. Chest computed tomography (CT) disclosed diffuse, thin-walled, and rounded lung cysts in bilateral lungs as shown in Fig. 1. Afterward, she experienced multiple episodes of chest pain when deep breath, dyspnea on exertion, and dry cough. Last year, spirometry demonstrating severe obstructive ventilatory defects. Follow up chest CT in 2020 showed diffuse, thin-walled, and rounded lung cysts in both lungs exaggerating, as shown in Fig. 2. Besides, gum-like sputum (proven chyloptysis later) has been noted since 2020/08/08, as shown in Fig. 3. Lymphangioleiomyomatosis was diagnosed and sirolimus has been prescribed for lung lymphangioleiomyomatosis since 2020/10/11. Under the sirolimus control, lung function test (LFT) showed improving forced expiratory volume in one second (FEV1) and forced vital capacity (FVC), as shown in Table 1. Meanwhile, gum-like sputum decreased, and dyspnea subsided with time.

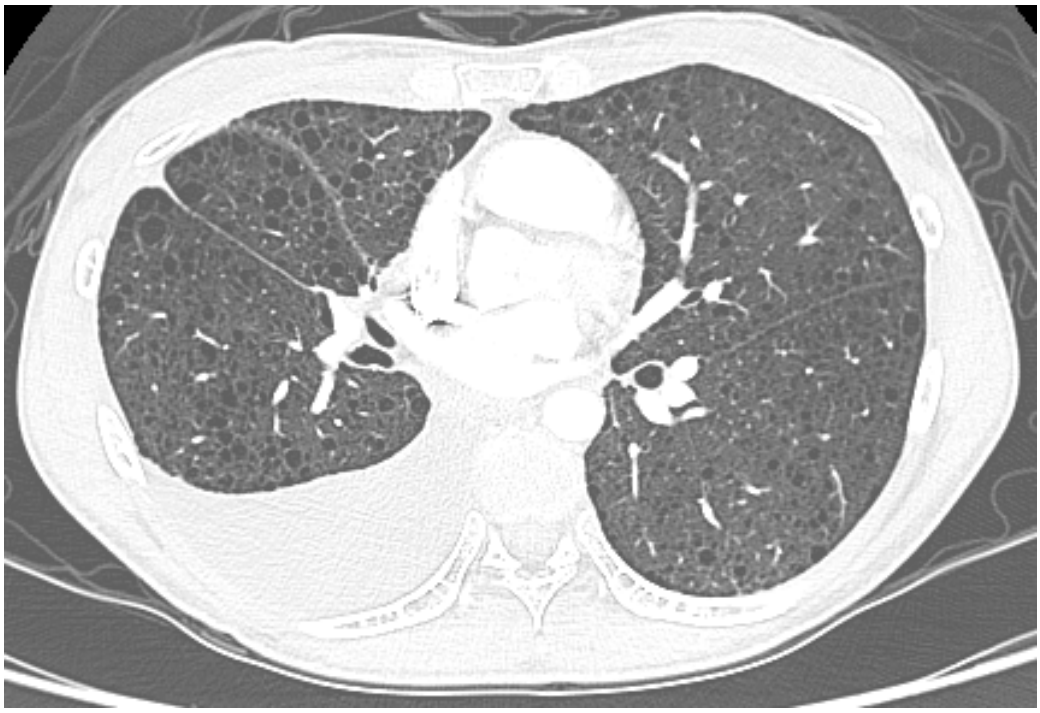

Fig. 1 Chest CT on 2012/09/18 disclosed 1. R't pleural effusion (proven chylothorax later) and 2. diffuse, thin-walled, and rounded lung cysts in bilateral lungs.

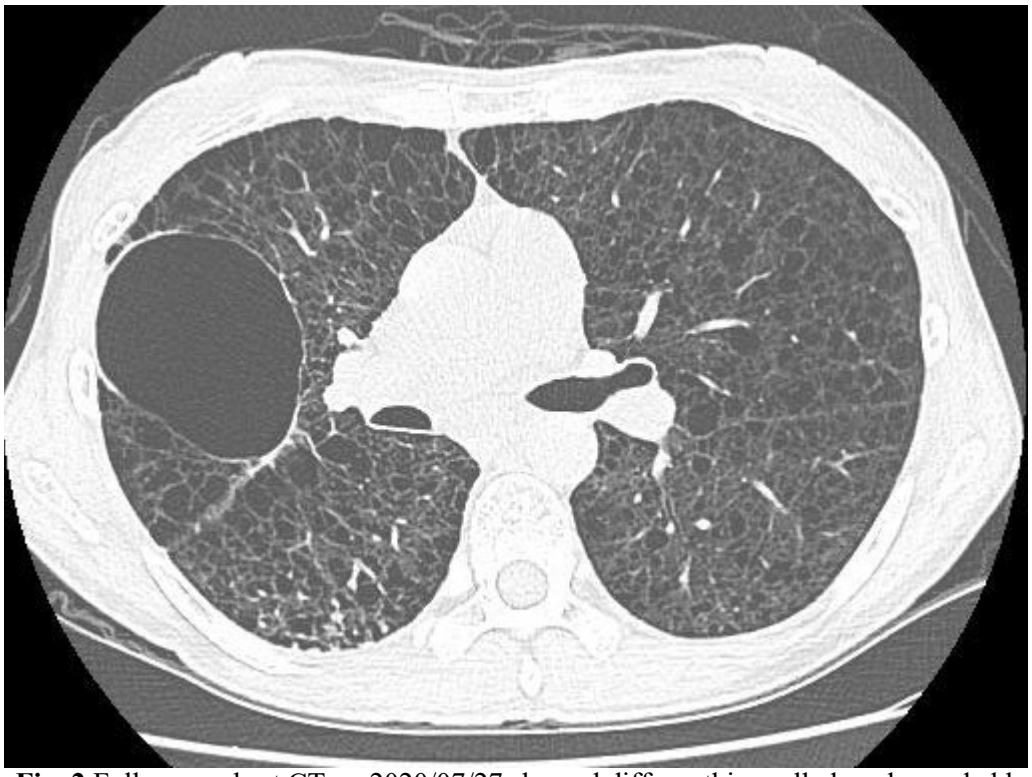

Fig. 2 Follow up chest CT on 2020/07/27 showed diffuse, thin-walled, and rounded lung cysts in both lungs exaggerating

Fig. 3. Gum-like sputum (proven chyloptysis later)

Date	FEV1	FVC	FEV1/FVC
2012/09/20	2.06 L	2.58 L	80%
2020/03/11	1.06 L	1.83 L	58%
2020/09/05	1.20 L	2.06 L	58%
2020/11/07	1.37 L	1.91 L	72%
2021/01/09	1.83 L	2.61 L	70%

Table 1. Lung function test (LFT) showed improving forced expiratory volume in one second (FEV1) and forced vital capacity (FVC).

Discussion

Chyloptysis is a rare symptom and the differential diagnosis of chyloptysis includes lymphangioleiomyomatosis, yellow nail syndrome, or thoracic lymphangiectasis. [1] Lymphangioleiomyomatosis (LAM) is a rare, progressive, cystic lung disease that occurs primarily in women of child-bearing age. It can occur sporadically (S-LAM) or in conjunction with tuberous sclerosis complex (TSC-LAM), an autosomal dominant. [2] Our case belongs to sporadic lymphangioleiomyomatosis (S-LAM) with the prevalence of 1 to 5 per million. [3, 4] The most common manifestations of LAM were pneumothorax and dyspnea on exertion, and less common presentations were chylothorax, hemoptysis, and ascites. [5,6] Chyloptysis was rare with the incidence of 3%, and besides chyloptysis, the rare presentation of LAM included chylous ascites and chyluria. [5,6] A lymphangiogram which could demonstrated a direct communication between the thoracic duct and lymphatic space was able to show the lymphatic efflux into the bronchi. [7] Sirolimus inhibits mTOR and improved the forced expiratory volume in 1 second (FEV_1) in the first year. [8] Pulmonary function was maintained and stabilized up to 4 years. [8] In sight of the clinical presentation and treatment result, our case was consistent with the study.

Conclusion

Chyloptysis is a rare symptom, and the differential diagnosis includes lymphangioleiomyomatosis (LAM). Sirolimus could improve pulmonary function in the patients with LAM.

References

1. Lim KG, Rosenow EC 3rd, Staats B, Couture C, Morgenthaler TI. Chyloptysis in adults: presentation, recognition, and differential diagnosis. Chest. 2004 Jan;125(1):336-40. doi: 10.1378/chest.125.1.336. PMID: 14718464.
2. Steagall WK, Pacheco-Rodriguez G, Darling TN, Torre O, Harari S, Moss J. The Lymphangioleiomyomatosis Lung Cell and Its Human Cell Models. Am J Respir Cell Mol Biol. 2018 Jun;58(6):678-683. doi: 10.1165/rcmb.2017-0403TR. PMID: 29406787; PMCID: PMC6002654.
3. Johnson SR. Lymphangioleiomyomatosis. Eur Respir J. 2006 May;27(5):1056-65. doi: 10.1183/09031936.06.00113303. PMID: 16707400.
4. McCormack FX. Lymphangioleiomyomatosis. MedGenMed. 2006 Jan 18;8(1):15. PMID: 16915145; PMCID: PMC1682009.
5. Taylor JR, Ryu J, Colby TV, Raffin TA. Lymphangioleiomyomatosis. Clinical course in 32 patients. N Engl J Med. 1990 Nov 1;323(18):1254-60. doi: 10.1056/NEJM199011013231807. PMID: 2215609.
6. Chu SC, Horiba K, Usuki J, Avila NA, Chen CC, Travis WD, Ferrans VJ, Moss J. Comprehensive evaluation of 35 patients with lymphangioleiomyomatosis. Chest. 1999 Apr;115(4):1041-52. doi: 10.1378/chest.115.4.1041. PMID: 10208206.
7. Lim KG, Rosenow EC 3rd, Staats B, Couture C, Morgenthaler TI. Chyloptysis in adults: presentation, recognition, and differential diagnosis. Chest. 2004 Jan;125(1):336-40. doi: 10.1378/chest.125.1.336. PMID: 14718464.
8. Hu S, Wu X, Xu W, Tian X, Yang Y, Wang ST, Liu S, Xu X, Xu KF. Long-term efficacy and safety of sirolimus therapy in patients with lymphangioleiomyomatosis. Orphanet J Rare Dis. 2019 Aug 20;14(1):206. doi: 10.1186/s13023-019-1178-2. PMID: 31429781; PMCID: PMC6702727.