

INTRODUCTION

- Pulmonary lymphangitic carcinomatosis is a rare process in which cancer cells spread to the lymphatic system of the lungs causing obstruction.
- The first case of "lymphangitis carcinomatosa" was described by Andral et al in 1824 secondary to uterine cancer.
- PLC carries a high mortality, approximately 50% of patients die within 2 months of first respiratory symptoms.
- Adenocarcinoma is the malignancy most commonly associated with LC. Others include breast, renal cell, and many others.

BRIEF SUMMARY OF CASE

- 68 year male presents with a three month history of progressive dyspnea on exertion.
- He had a contrasted CT of his chest performed three months prior to presentation and noted were innumerable scattered bilateral pulmonary nodules – largest measuring 12 mm. He was lost to follow up.
- A CT guided biopsy was performed after which he suffered PEA arrest. ROSC was achieved though patient died four days later.
- Pathology eventually demonstrated renal cell carcinoma.

OBJECTIVES

- Describe the epidemiology, physiology, and diagnosis for lymphangitic carcinomatosis.
- Identify the most common signs and symptoms
- Describe imaging findings associated with PLC
- Understand the most common malignancies associated with lymphangitic carcinomatosis

HISTORY AND CLINICAL COURSE

Mr. B was a functional 68-year-old male who presented to the ED with a chief complaint of progressive dyspnea on exertion associated with dry cough over the last ten weeks. Other pertinent symptoms included profound orthopnea, poor appetite with nausea and a steady 20lb weight loss over the preceding three months. He denied hemoptysis, night sweats, fevers or chills.

He had a contrasted CT of his chest performed three months prior to presentation and incidentally noted were innumerable scattered bilateral pulmonary nodules – largest measuring 10-12 mm. He was then lost to follow up.

Distant family history on presentation was notable for exposure to pulmonary tuberculosis as a child – grandfather with active pulmonary TB for >10 years.

He was a guitar player for a local band. He had no known asbestos or occupational exposures. Though he denied history of smoking, he often played music in venues where he was exposed to second hand smoke. He rarely drank alcohol and denied use of illegal drugs.

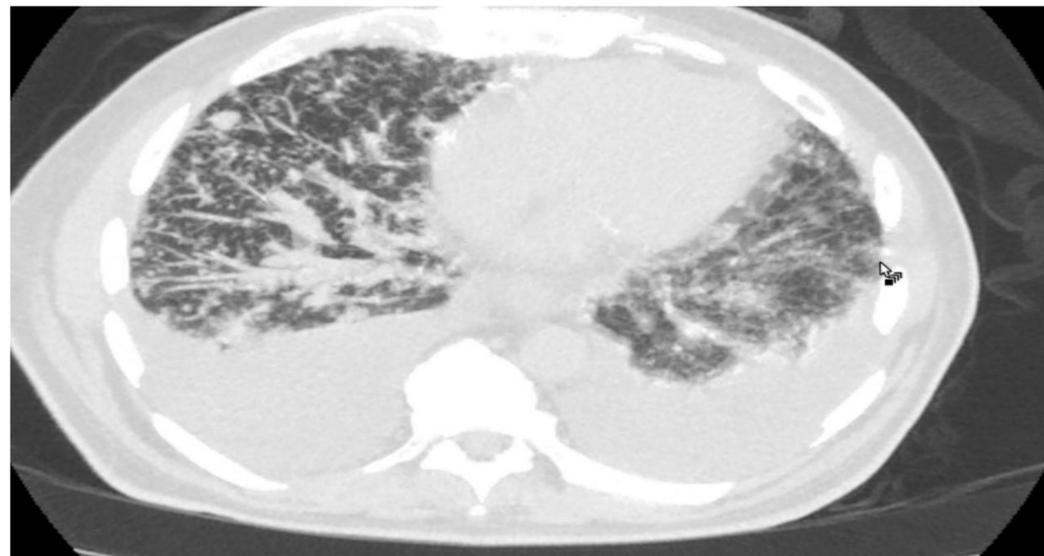
Objective:

- Notable pulmonary findings of mild respiratory distress, bibasilar rales, and dullness to percussion at his bases. Additionally, there was mild tenderness to palpation over his epigastrium.
- Labs showed mildly elevated Calcium of 10.4 which corrected to 10.7. The remainder of his complete blood count and metabolic panel were within normal limits. Additional studies included a mildly elevated CRP 1.02, IgE Ag elevated 2084.
- A QuantiFERON gold was obtained on which later resulted negative. COVID-19 also came back negative.
- High resolution CT of his chest obtained during his initial work-up demonstrated diffuse lung nodules with interval progression from prior which followed a lymphangitic distribution and with a bi-basal predominance.

A right sided dominant nodule was thought to be challenging for CT guided biopsy. Given its location at the right middle fissure, a thoracentesis was first performed. No malignant cells or bacteria were identified, and cytology was non-specific. Patient was eventually sent for a CT guided biopsy of dominant right middle fissure 1.2 cm nodule and tissue was successfully obtained.

Shortly after biopsy the patient suffered PEA of unknown etiology. ROSC was achieved after 40 minutes of CPR. He continued to deteriorate after arrest and on post arrest day 4 he was transitioned to comfort care. He passed away less than 24 hours later. Pathology from biopsy showed metastatic renal cell carcinoma

IMAGING



DISCUSSION

Lymphangitic carcinomatosis is a rare process in which cancer cells spread to the lymphatic system causing obstruction. Most commonly occurs in the lungs, called pulmonary lymphangitis carcinomatosis (PLC) and most commonly occurs with adenocarcinomas of the breast, lung, colon, stomach, pancreas and prostate. In a literature review renal cancer was found in 7.9% of PLC. Most common symptoms of dyspnea and dry cough are nonspecific and often lead to missed or delayed diagnosis. It typically carries with it a generally poor prognosis with diagnosis often made postmortem. Diagnostic consideration for lymphangitic carcinomatosis should be in patients with unexplained hypoxemia and progressive dyspnea with known or high suspicion for malignancy. Initial differential diagnosis for this patient included disseminated pulmonary tuberculosis, sarcoidosis, silicosis (no known exposures), vasculitis, metastatic cancer.

The mechanism of metastatic spread is not exactly known. One hypothesis states that malignant cells may undergo hematogenous spread to the lungs where lymphatic invasion may occur. Another consideration is retrograde metastasis into lymphatics from the mediastinal and hilar lymph nodes, though this would be more likely associated with spread from organs that share the thoracic lymph system. What we do know is that not only are lymphatics involved on histological examination, but the adjacent interstitium also shows evidence of tumor on histologic examination with associated edema and desmoplasia.

HRCT is the preferred imaging modality. Findings include interlobular septal thickening resulting in prominent definition of the secondary pulmonary lobules which look like "tessellating polygons". The combination of interlobular septal thickening from lymphangitis along with the prominence of the central lobular bronchovascular bundle creates a "dot in box" appearance - typically in a non-dependent pattern. There is often significant hilar or mediastinal nodal enlargement along with overall preservation of the lung architecture.

Lung biopsy is recommended for confirmation, though diagnosis can be made based on imaging and clinical presentation.

REFERENCES

- Biswas, Abhishek and Sriram, Peruvemba. "Getting the Whole Picture: Lymphangitic Carcinomatosis". *The American Journal of Medicine*. Apr 22 2015. Vol 128, Iss 8, P837-840. [https://www.amjmed.com/article/S0002-9343\(15\)00347-2/fulltext](https://www.amjmed.com/article/S0002-9343(15)00347-2/fulltext)
- Gudbjartsson T, Thoroddsen A, Petursdottir V, et al. Effect of incidental detection for survival of patients with renal cell carcinoma: results of population-based study of 701 patients. *Urology* 2005; 66:1186.
- Guddati A, KGuddati A, K, Marak C, P: Pulmonary Lymphangitic Carcinomatosis due to Renal Cell Carcinoma. *Case Rep Oncol* 2012;5:246-252. doi: 10.1159/000339126
- Johkoh T, Ikezoe J, Tomiyama N et-al. CT findings in lymphangitic carcinomatosis of the lung: correlation with histologic findings and pulmonary function tests. *AJR Am J Roentgenol*. 1992;158 (6): 1217-22. *AJR Am J Roentgenol* (abstract) - Pubmed citation
- Klimek, M (2019) Pulmonary lymphangitis carcinomatosis: systematic review and meta-analysis of case reports, 1970-2018, *Postgraduate Medicine*, 131:5, 309-318, DOI: 10.1080/00325481.2019.1595982
- Lin WR, Lai RS. Pulmonary lymphangitic carcinomatosis. (2014) *QJM : monthly journal of the Association of Physicians*. 107 (11): 935-6. doi:10.1093/qjmed/hcu076 - Pubmed