



## Case report

# Transudative chylothorax in a patient with lymphangioleiomyomatosis



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## ABSTRACT

Transudative chylothorax is a rare type of pleural effusion. It has been described to be present in the setting of liver cirrhosis, heart failure, amyloidosis, nephrotic syndrome, superior vena cava thrombosis, sclerosing mesenteritis and heart failure.

We present the case of an 86-year-old woman with a right-side transudative chylothorax associated with heart failure and lymphangioleiomyomatosis.

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## 1. Introduction

Pulmonary lymphangioleiomyomatosis (LAM) is a rare systemic disease in mainly in woman, characterized by an abnormal smooth muscle proliferation in the peribronchial, perivascular and perilymphatic regions of the lung. It occurs sporadically or associated with tuberous sclerosis complex.

Chylothorax is a well described complication of LAM and usually results from obstruction or disruption of the thoracic duct [1].

The diagnosis of chylothorax is usually pursued when the appearance of the fluid is “milky” or when thoracic duct injury is suspected to be the cause of a pleural effusion [1].

Chylous effusions are typically exudates high in triglycerides and chylomicrons. However, in a retrospective series of 74 patients, less than 15% were transudates [1]. Chylothorax can present as a transudate when associated to other concomitant conditions like: cirrhosis, amyloidosis, nephrotic syndrome, superior vena cava thrombosis, sclerosing mesenteritis and congestive heart failure [3–7]. Here we present a case of patient with LAM with recurrent transudative chylothorax.

## 2. Case description

An 86 year-old Caucasian woman, presented with 4 weeks of

progressive worsening of shortness of breath. Her past medical history is remarkable for a recent pulmonary embolism with right transudative pleural effusion, lung cysts and renal angiomyolipomas. Her physical exam was remarkable for diminished breath sounds on auscultation of the right lung base, multiple skin hyperchromic papules and bilateral 2 + pitting edema up to her knees. The remainder of her exam was normal. Chest roentgenogram done on admission showed a large opacification of the right hemithorax suggestive of a pleural effusion. Thoracentesis was performed and 1200 ml of cloudy yellowish fluid was drained. Pleural fluid analysis revealed a cell count with 1231 WBC, with 97% lymphocytes, pleural albumin 1.5 mg/dL, serum albumin 3.2 mg/dL, pleural LDH 89 mg/dL, serum LD 234 mg/dL, triglycerides 205 mg/dL, cholesterol 49 mg/dL. The fluid analysis confirmed the presence of a transudative effusion by light's criteria, with lymphocytic predominance. Also the high triglyceride and low cholesterol level confirmed a chylothorax. Cytology and microbiologic studies were unremarkable Figs. 1–3.

An echocardiogram done on admission revealed a preserved ejection fraction with grade 1 diastolic dysfunction and increased right ventricular systolic pressure of 81 mmHg, with mildly dilated right ventricle size.

Computed tomography done after thoracentesis showed again the presence of multiple lung cysts, which were seen in previous imaging studies, with minimal residual pleural effusion on the right hemithorax.

Patient persisted with recurrent pleural effusion and required evaluation by cardiothoracic surgeon, who performed wedge resection in the right upper lobe and talc pleurodesis with success. Pathology showed areas of smooth muscle hyperplasia and

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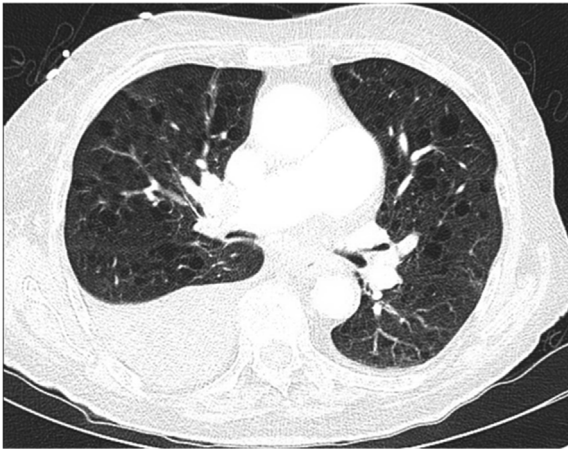


Fig. 1. CT chest axial view, showing right pleural effusion and multiple lung cysts.

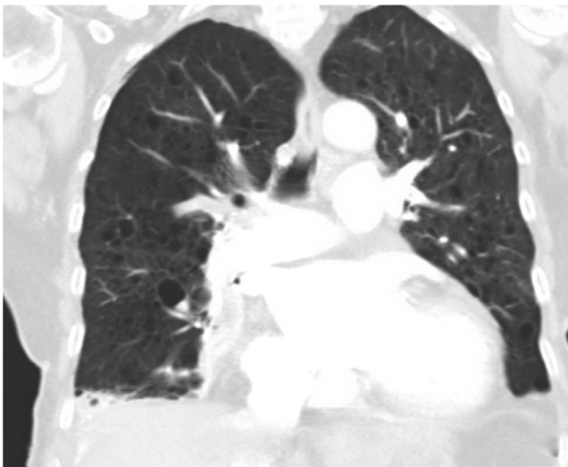


Fig. 2. CT chest coronal view, showing bilateral diffuse lung cysts.

immunostains were positive for smooth muscle actin, podoplanin, progesterone and estrogen receptor confirming diagnosis of tuberous sclerosis associated LAM.

On further follow up after 1 year; the patient has stable minimal right pleural effusion and improved shortness of breath. She has not required subsequent thoracentesis since then.

### 3. Discussion

Chylous pleural effusions are typically described as exudative lymphocytic pleural fluid with a milky appearance, however less than half of chylothorax have a milky appearance [1].

The causes of chylothorax may be divided into four major categories: tumor, trauma, idiopathic, and miscellaneous [8].

Pleural fluid from a chylothorax usually has a triglyceride level above 110 mg/dL, low cholesterol level, and elevated lymphocyte count. Also a ratio of the pleural fluid to serum triglyceride level of greater than 1.0, and a ratio of the pleural fluid to serum cholesterol level of less than 1.0 has been described [2]. Moreover, the presence of chylomicrons in lipoprotein analysis of the pleural fluid confirms the diagnosis of chylothorax. This is particularly valuable if the triglyceride level is in the ranges of 50–110 mg/dL [9].

Chylothorax are usually exudative effusion with milky appearance. Because triglyceride levels also vary in patients with



Fig. 3. Cloudy yellow pleural fluid immediately after thoracentesis.

chylothorax, traditional triglyceride cutoff values used in excluding the presence of chylothorax may miss the diagnosis in fasting patients, particularly in the postoperative state [1].

The management of chylothorax in the setting of LAM involves pleural drainage, pleurodesis thoracic duct ligation, pleurectomy, low fat diet, and sirolimus.

Conservative management with diet is usually unsuccessful. Sirolimus is a mammalian target of rapamycin (mTOR) inhibitor. In many institutions is the 1st line of treatment due to 2 observational studies that assessed the role of sirolimus for the management of chylothorax associated to LAM showing that it decreases the size of the effusion and in some cases it can resolve the entire chylothorax [10,11].

More invasive options like pleurodesis or thoracic duct ligation are used when more conservative approaches do not respond.

The presence of heart failure with preserved ejection fraction and pulmonary hypertension most likely due to her recent pulmonary embolism, may have contributed to the presentation of a transudative effusion in this case. Also because of her effusion being most likely multifactorial it was decided to have a more aggressive approach first.

Finally chylothoraces may present with diverse pleural fluid appearance and biochemical characteristics. Approximately 20 cases of transudative pleural effusion have been described in the setting of chylothorax, many times diagnosis is missed because pleural fluid does not present with a milky appearance or it is a transudate fluid like in this case, therefore pleural triglyceride and cholesterol levels should be ordered whenever you suspect of chylothorax. To our knowledge this is the first case reported of transudative chylothorax in the setting of LAM.

This case emphasizes that is important to be aware of the variety of presentations of chylothorax to ensure an early diagnosis and treatment of this condition.

### Conflict of interest

None declared.

### Authorship statement

Adrian Noriega Aldave contributed to this case report by writing the paper.

John W Leslie Jr contributed to this case report by revising and correcting the manuscript.

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