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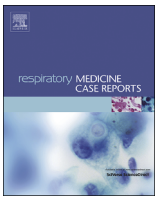

Case Report

Refractory chylothorax in a patient with Waldenström Macroglobulinemia: A case report

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A B S T R A C T

A refractory chylothorax secondary to Waldenström Macroglobulinemia (WM) has rarely been reported in literature, but often responds to chemotherapy and conservative measures. Few reports have been published reporting the use of surgical intervention when standard medical therapies fail. We present a 76-year-old male who developed a large right sided chylothorax with a soft tissue mass encasing the descending thoracic aorta. Pleural fluid flow cytometry and biopsy of the mass demonstrated findings diagnostic of WM. Despite chemoimmunotherapy and conservative measures, he required frequent, high-volume, therapeutic thoracentesis for relief of dyspnea. Thoracic duct embolization (TDE) was performed which resolved the patient's chylothorax, however he subsequently developed lower volume serosanguinous pleural effusions. Patient continued requiring therapeutic thoracentesis bimonthly and ultimately proceeded with pleurectomy of the right lung to achieve resolution of symptoms. To our knowledge, this is the first reported case where pleurectomy has successfully treated refractory pleural effusions in a patient with WM.

1. Introduction

A chylothorax is caused when chyle enters the pleural cavity by traumatic or non-traumatic means. Non-traumatic chylothoraxes are uncommon causes of exudative pleural effusions and can be observed as a complication of malignancy; however, Waldenström Macroglobulinemia (WM) has rarely been described as a potential cause. A recurring chylothorax causes loss of essential proteins, immunoglobins, fats, vitamins, and electrolytes which can lead to hypovolemia and severe malnutrition [1]. Frequent therapeutic thoracentesis can provide relief in respiratory symptoms, however, malnutrition from chyle loss will persist and progress if the leakage is not fixed [1]. Few cases of a reoccurring chylothorax secondary to WM have been reported that resolved with medical therapy and conservative measures, however, cases using surgical interventions are limited [2]. Literature review has not demonstrated the use of pleurectomy as a potential treatment for patients with a refractory chylothorax in patients with WM who have failed medical therapy and conservative measures. We present a patient with WM and a recurring chylothorax refractory to chemotherapy and conservative measures, who underwent thoracic duct embolization (TDE) and eventual pleurectomy to achieve resolution of symptoms.

2. Case presentation

A 76-year-old male former smoker with a significant history of hypertension presented to his outpatient primary care office for a dry cough (Fig. 1). Computed tomography (CT) of the chest demonstrated a large right-sided pleural effusion with suggestion of subcarinal lymphadenopathy. The patient was evaluated by a pulmonologist and underwent a right-sided thoracentesis. Two liters (L) of pleural fluid appearing milky-orange was drained (Fig. 2). Pleural fluid studies demonstrated a triglyceride concentration of 410 mg/dL confirming the presence of a chylothorax. Additional contrasted CT of the chest showed an ill-defined soft tissue mass encasing the

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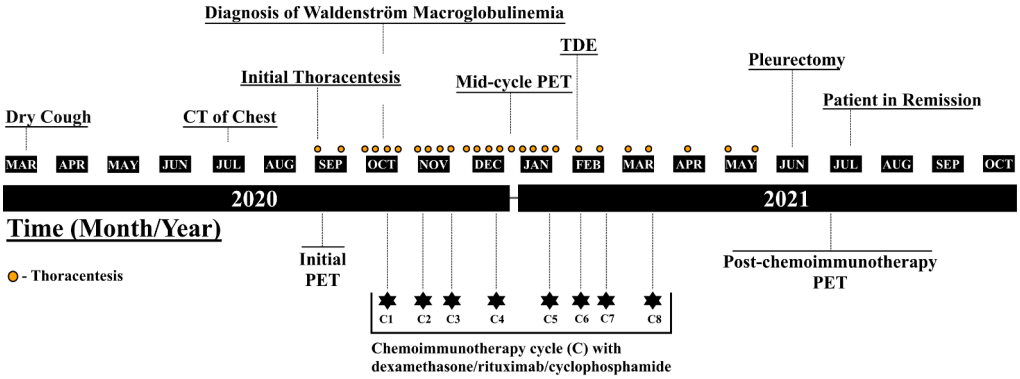

Fig. 1. Abbreviated timeline of clinical course demonstrating the time of symptom onset to the time of remission.

Fig. 2. Pleural fluid from initial thoracentesis appearing milky-orange in color. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

descending thoracic aorta (Fig. 3). Follow up positron emission tomography (PET) demonstrated abnormal FDG avidity of the soft tissue mass (Fig. 3). Tissue sampling via endobronchial ultrasound (EBUS) demonstrated a monomorphic population of small lymphocytes expressing CD19, CD20, and CD22 while being negative for CD5 or CD10. Further oncological evaluation including the detection of a MYD88 L265P mutation led to a diagnosis of WM. The patient was started on combination chemoimmunotherapy with cyclophosphamide, dexamethasone, and rituximab (Fig. 1). Despite excellent radiologic response to chemoimmunotherapy (Fig. 3), he continued requiring high volume therapeutic thoracentesis (2.0–2.5 L per week). This ultimately led to 26 therapeutic thoracenteses leading to weight loss and malnutrition (Fig. 1). The patient was referred to an interventional radiologist with expertise in lymphatics and underwent TDE. Within weeks of successful TDE, the patient's milky pleural effusion transitioned to a serosanguinous lower-

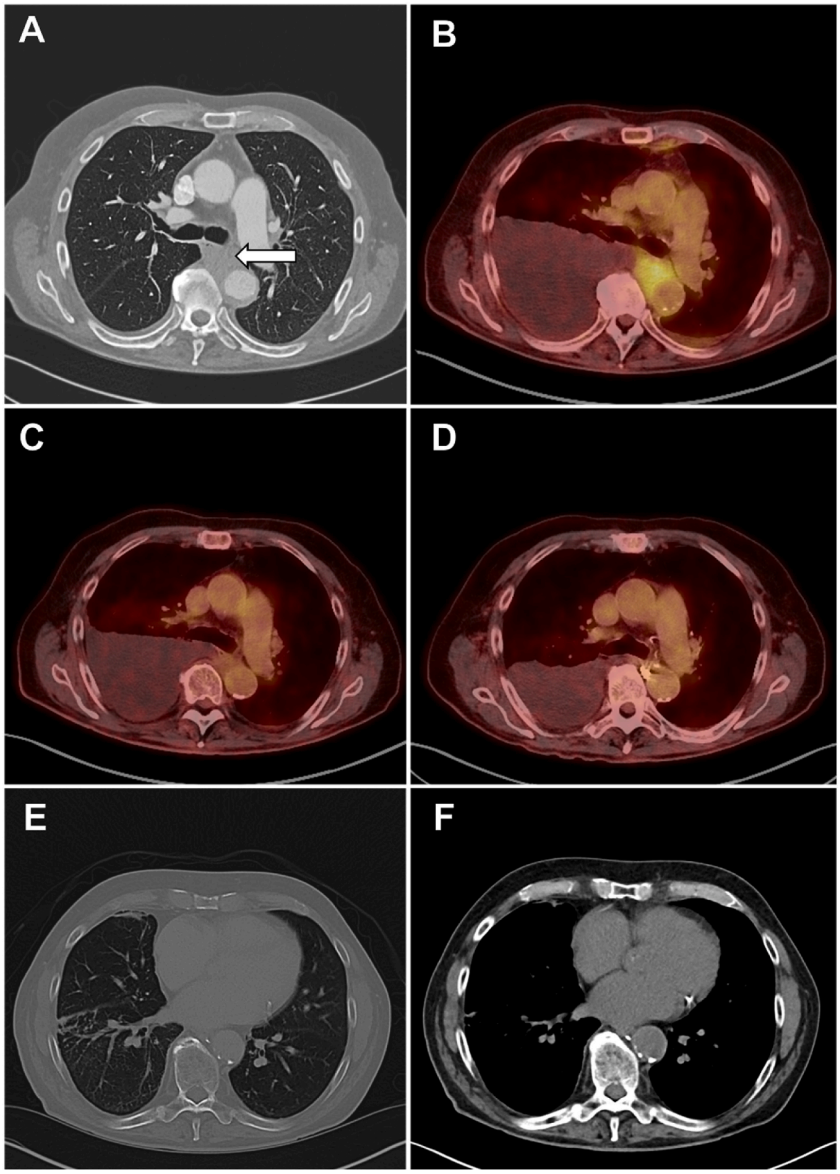

Fig. 3. (A) Contrasted CT of the chest demonstrating a periaortic mass (white arrow). (B) Initial PET demonstrating abnormal FDG-avidity of the soft mass encasing the descending thoracic aorta. (C) PET mid-chemoimmunotherapy demonstrating interval decrease in size and FDG uptake of the soft tissue in the posterior mediastinum. (D) PET post-chemoimmunotherapy demonstrating no evidence of disease or FDG-avidity of the soft mass encasing the descending thoracic aorta. CT of the chest post pleurectomy demonstrating resolved pleural effusion; (E) lung enhanced view and (F) mediastinal view.

volume pleural effusion with a triglyceride concentration of 60 mg/dL. The patient, however, continued requiring therapeutic thoracentesis bimonthly for these serosanguinous appearing pleural effusions (1.0–1.5 L bimonthly). He was referred to thoracic surgery for further evaluation, and ultimately underwent a successful pleurectomy of the right lung. Follow-up pleural ultrasound four weeks post procedure demonstrated <50 ml of pleural fluid indicating excellent response to surgical intervention (Fig. 3). Patient completed chemoimmunotherapy without evidence of disease or FDG avidity on repeat PET (Fig. 3), and is currently in remission for his WM. He reports significant improvements in quality-of-life with resolution of dyspnea and has not required further therapeutic thoracentesis since his pleurectomy.

3. Discussion

In cases of individuals who have been diagnosed with WM, only a handful have reported chylothorax as a complication [2]. Of those cases reviewed, WM associated with a recurring chylothorax commonly improve with chemotherapy and conservative measures [2,3]. Our patient, however, continued requiring weekly large-volume therapeutic thoracentesis despite months of effective chemoimmunotherapy. In refractory cases such as ours, TDE can be performed for chylous leaks and has a reported success rate of

73–90% with minimal complications [4]. While TDE technically treated this patients' refractory chylothorax as evidenced by a normalized pleural triglyceride content, he developed a subsequent recurrent serosanguinous appearing pleural effusion. Further intervention was necessary, and patient was referred to thoracic surgery for surgical intervention. After video-assisted thoracoscopy demonstrated a thickened rind on the majority of the parietal pleura, pleurectomy was performed over pleurodesis. Patient has not required a therapeutic thoracentesis since his pleurectomy. In review of current literature, only one other case reports the use of pleurectomy for managing a refractory chylothorax in WM, but was ultimately reported unsuccessful [5]. Here we report the first successful use of pleurectomy for a refractory chylothorax in WM. In individuals with refractory chylothorax secondary to malignancy, a step wise approach is necessary, and only surgical interventions such as TDE and pleurectomy should be performed if chemotherapy and conservative measures fail.

4. Conclusion

A chylothorax in patients with malignancies such as Waldenström macroglobulinemia often improve after chemotherapy and conservative measures. In cases refractory to medical therapy, surgical intervention such as TDE, pleurodesis, or pleurectomy should be considered.

Declaration of competing interest

The authors have no conflicts of interests to declare.

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