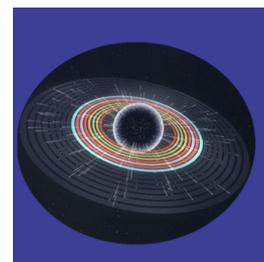


WW Med 2019 Vol 1 issue: 6 214-216

Worldwide Medicine



Case Report

Refractory Chylothorax ; A rare presentation of Systemic Lupus Erythematosus (SLE)

Adrian Mark Masnammany, Asmahan Mohamad

Rheumatology Unit, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia.

Received: 09 August 2019 / Accepted: 14 August 2019

Abstract

5-10% of SLE patients can present with pleuritis as their initial manifestation. However, chylothorax is a rare finding in patients with SLE. A 27 year old lady presented with refractory bilateral chylothorax of unknown etiology and was later diagnosed as SLE. Her chylothorax resolved with immunosuppressive therapy, however had a recurring course throughout her follow up.

Keywords: serositis, chylothorax, effusion, sle, refractory

Introduction

Pulmonary involvement is seen in 30-50% of SLE patients, including pleuritis, pleural effusion, pneumonitis, interstitial lung disease and pulmonary hypertension . SLE is the most common connective tissue disease with pleural involvement. However, chylothorax is an exceedingly rare complication seen in SLE, and based on literature search, only 7 cases have been reported. We describe a case of SLE with initial presentation of chylothorax.

Case Summary

A 27year old lady of malay descent, with previously no known medical illness presented with history of progressive breathlessness of 1 week duration. She had no history of fever, cough , weight loss or other constitutional symptoms. No history of orthopnoea or paroxysmal nocturnal dyspnoea. She had no symptoms or prior history suggestive of connective tissue disease. No history of trauma or previous thoracic surgery. Clinically she was tachypnoeic and lung

Corresponding Author: Adrian Mark Masnammany - Rheumatology Unit, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia.; email: adrianmark876@gmail.com

DOI: 10.5455/ww.59800

This is an Open Access article under the terms of the Creative Commons Attribution-Non-Commercial 4.0 International License (<https://creativecommons.org/licenses/by-nc/4.0/>)

examination had reduced breath sounds bilaterally with stony dullness on percussion. She had no lymphadenopathy. Other examination findings were unremarkable. Chest radiograph had blunted costophrenic angles with meniscus sign, suggestive of bilateral large pleural effusion. She had bilateral chest tube inserted and drained large volumes odorless milky fluid. Pleural fluid analysis was exudative in nature based on Light's criteria, with elevated lactate dehydrogenase, cholesterol and triglyceride, suggestive of chylothorax. Gram stain, pleural fluid culture, acid fast stain, TB gene-expert and mycobacterial culture were all negative. Pleural fluid cytology was negative for malignant cells.

Computed Tomography (CT) thorax demonstrated bilateral loculated residual hydropneumothorax, passive collapse consolidation of bilateral lower lobes, with no other parenchymal lesion or mediastinal lymphadenopathy. No pericardial effusion or ascites seen.

She had pleuroscopy done, which showed multiple adhesions, multiple whitish to yellowish nodular lesions and hyperaemic parietal pleura.

Bronchoscopy showed generalized hyperaemia, oedematous mucosa with prominent vessels at bronchial mucosa and no endobronchial lesion or obstruction seen.

She was initially treated empirically for smear negative pulmonary and pleural tuberculosis, however had no clinical improvement despite 4 months on anti-tubercular drugs, and was stopped.

A rheumatology consult was obtained in view of unexplained chylothorax. Immunological studies came out positive for both anti-dsDNA and ANA with titre of 1:320 homogenous, as well as hypocomplementaemia and positive SSA/Ro. Other ENA (Extractable nuclear antigens), rheumatoid factor and antiphospholipid study were negative.

The diagnosis of SLE with bilateral chylothorax was made and patient was started on hydroxychloroquine and pulsed with intravenous methylprednisolone 500mg daily for 3 days and was put on oral prednisolone 1mg/kg. Her chylothorax improved subsequently and she was discharged with tapering doses of prednisolone. Subsequent review showed resolution of chylothorax while on oral prednisolone.

She did not have any other manifestation of lupus during her 9 year follow up period. However, had recurring right sided effusion throughout 9 years, with multiple readmissions for symptomatic chylothorax requiring needle thoracocentesis and escalation of her prednisolone doses. She was allergic to azathioprine, and was started on cyclosporin as steroid sparing agent.

She was planned for indwelling tunnelled pleural catheter but postponed due to bouts of infection requiring multiple hospitalization. She subsequently developed septic shock secondary to severe pneumonia and succumbed.

DISCUSSION

SLE patients can present with serositis; either in the form of pleural effusion, ascites or pericardial effusion. The pleural effusion is usually bilateral, and can produce symptoms of breathlessness, pleuritic chest pain or even be asymptomatic. The fluid analysis is exudative in nature, may contain neutrophils or lymphocytes, low glucose levels and low complements. Our patient did not have ascites nor pericardial effusion.

In most patients, serositis responds well to glucocorticoids and may resolve without any sequelae or may run a recurrent course, such in the case of our patient. Occasionally patients may develop pleural thickening or complex effusion.

Chylothorax refers to the accumulation of chyle within the pleural space due to disruption of the thoracic duct or its divisions and clinically characterized by a milky odourless fluid containing high levels of triglycerides. The diagnosis of chylothorax is made based on pleural triglyceride levels greater than 110 mg/dl, (while a level <50 mg/dL excludes a chylous effusion) and the ratio of the pleural fluid to serum cholesterol is less than 1.0.

Common causes for chylothorax are mainly traumatic due to surgical procedure or chest trauma. Nontraumatic causes include malignancy, liver cirrhosis, nephrotic syndrome, congestive cardiac failure, lymphagioliomyomatosis and infections like filariasis, tuberculosis or histoplasmosis.

Tuberculosis is endemic in our country and was initially presumed to be the etiology for chylothorax in our patient. However, extensive work up for tuberculosis turned out negative including failed trial of empirical treatment with anti-tubercular regime.

A limited number of autoimmune diseases have been associated with chylothorax, albeit rare including sarcoidosis, behcet disease and SLE.

Strausser and Flye² described a 24-year old lady with underlying SLE complicated with a left-sided chylothorax and chylous ascites.

Lee et al.² reported two cases with previously undiagnosed SLE presenting as bilateral chylothorax, chylous ascites, and protein-losing enteropathy. However, these patients had concurrent proteinuria and malar rash prompting the diagnosis of SLE.

Y.J. Lin et al³ reported a 43year old previously healthy lady presenting with left sided chylothorax with lymphopenia, positive ANA, dsDNA and ENA.

Bao-Bao Hsu et al⁴ reported a 21year old girl with underlying SLE and refractory bilateral chylothorax not responsive to steroids and cyclophosphamide.

E.S Dilek et al⁵ reported a 61year old lady with underlying SLE for 10years, presenting with symptomatic chylous ascites and bilateral chylothorax and Basanta el al⁶ reported a 19year old girl with underlying SLE presenting with left sided chylothorax.

The diagnosis of chylothorax as a sole manifestation in SLE can be clinically challenging.

The exact pathophysiology for chylothorax in SLE is not well understood , and is likely inflammatory in nature.

A probable pathogenetic mechanism may include lymphatic vessel inflammation with resultant increased wall permeability , increased endoluminal pressure and extravasation of chyle into the pleural space.

Conclusion:

SLE patients can present with a multitude of clinical symptoms. Chylothorax is an unexpected finding in SLE. In patients presenting with unexplained nontraumatic chylothorax, after a careful exclusion of other etiologies, autoimmune diseases especially SLE need to be considered as a potential differential diagnosis. Although it may respond to glucocorticoid therapy, it may be refractory in some cases and pose a challenge in its management.

Conflict of interest

All authors declare that they have no conflict of interest

Funding

There was no funding received for this paper.

References:

1. Strausser JL, Flye MW (1981) Management of nontraumatic chylothorax. *Ann Thorac Surg* 31:520–526
2. Lee CK, Han JM, Lee KN et al (2002) Concurrent occurrence of chylothorax, chylous ascites, and protein-losing enteropathy in systemic lupus erythematosus. *J Rheumatol* 29:1330–1333
3. Y.-J. Lin , D.-Y. Chen , J.-L. Lan et al (2007) Chylothorax as the initial presentation of systemic lupus erythematosus: a case report. *Clin Rheumatol* (2007) 26:1373–1374
4. Bao Bao Hsu et al (2010) Refractory chylothorax in a patient with systemic lupus erythematosus: case report *Formosan Journal of Rheumatology* 2010;24:88-91
5. E.S Dilek (2013) A Rare Case of Systemic Lupus Erythematosus with Chylous Ascites and Chylothorax Case Report *Rheumatology* 2013; 2013: 797696.
6. Basanta et al (2015) Chylothorax in a case of systemic lupus erythematosus : a case report *The Pulmo-Face*; 2015, 15:1, 31-33)