

A rare case of nephrotic syndrome with chylothorax

Saurabh K. Singh, Ajay Chauhan, Bishakha Swain

Department of Medicine, ABVIMS Dr. RML Hospital, New Delhi, India

ABSTRACT

Chylothorax is an infrequent cause of pleural effusion that is most commonly caused by the obstruction or disruption of the thoracic duct. Chylothorax is rare in nephrotic syndrome. Unilateral chylothorax of the right side is due to the transdiaphragmatic shunting of chylous ascites. It is usually transient and self-limiting but a massive chylothorax requiring therapeutic thoracentesis can also be encountered. Here, we present a rare cause of chylous ascites-nephrotic syndrome resulting in chylothorax, where initially therapeutic thoracentesis is done followed by the management of nephrotic syndrome with modified Ponticelli regimen. This case highlights the need to consider chylous ascites as a cause of chylothorax via transdiaphragmatic shunting in patients with nephrotic syndrome to institute the appropriate treatment.

Keywords: Chylothorax, chylous ascites, nephrotic syndrome, transdiaphragmatic shunting, lymphangiography

Introduction

The nephrotic syndrome presents with heavy proteinuria, hypoalbuminemia, hypercholesterolemia, edema, and hypertension. Membranous nephropathy is amongst the commonest causes of primary nephrotic syndrome in adults.^[1]

Chylothorax is characterized by milky and opalescent fluid in the pleural space. It is caused by tumor (lymphoma, malignancies), trauma, and TB.

Chylous ascites is a well-known complication of severe nephrotic syndrome. However, the coexistence of chylous ascites and chylothorax is rarely reported in the adult nephrotic syndrome.^[2] Chylous effusion is typically exudative but in a minority of patients, chylothorax may be transudative like in nephrotic syndrome.

The pathogenesis of chylous ascites in nephrotic syndrome is unknown but hypoalbuminemia-induced bowel edema may be a predisposing factor. Chylothorax can occur because of chylous ascites as the negative intrapleural pressure draws chyle through the diaphragmatic defects into the pleural space.^[3]

Case Report

A 32-year-old female from Delhi, known case of hypothyroidism for 3 years and without any other comorbidities, recently diagnosed as a case of nephrotic syndrome whose cause was under evaluation (renal biopsy report awaited) presented with abdominal distension, generalized body swelling, and shortness of breath for 5 days associated with right-sided chest pain that increased on inspiration was non-radiating and sharpshooting in character.

She was afebrile and her BP was 100/60 mm Hg, PR: 85/min. She was tachypneic (30/min), SPO₂: 88% RA and 97% on 6L O₂/min, pallor and bilateral pitting pedal edema were present, chest: absent breath sound in the right side of the chest, abdomen: distended with shifting dullness.

Address for correspondence: Dr. Ajay Chauhan, Professor of Medicine, R. No 104, Academic Block; ABVIMS, Dr. Ram Manohar Lohia Hospital; Ministry of Health and Family Welfare, Govt. of India, New Delhi - 110 001, India. E-mail: dr.ajay@rmlh.nic.in

Received: 30-12-2020

Revised: 11-06-2021

Accepted: 20-06-2021

Published: 30-09-2021

Access this article online

Quick Response Code:



Website:
www.jfmpc.com

DOI:
10.4103/jfmpc.jfmpc_2605_20

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Singh SK, Chauhan A, Swain B. A rare case of nephrotic syndrome with chylothorax. J Family Med Prim Care 2021;10:3498-501.

Therapeutic thoracentesis was done and pleural fluid studies were sent. The pleural fluid was milky white in appearance as shown in Figure 1. Ascitic fluid paracentesis was also done which was also milky white.

There was no history of recent surgery, catheter insertion, or trauma.

Chest X-ray showed (1) right pleural effusion as shown in Figure 2. The coronal Contrast enhanced computed tomography scan chest showed gross right-sided pleural effusion as shown in Figure 3.

Biochemical characteristics of pleural and ascetic fluid of the patient		
	Pleural fluid	Ascitic fluid
Cytology	25-50 lymphocytes	60-80 lymphocytes
Sugar	138 mg/dL	125
Protein/albumin	0.7 gm/0.2 gm	0.5 gm/0.3 gm
Triglycerides	172 mg/dL	202 mg/dL
Cholesterol	80 mg/dL	67
LDH	158 mg/dL	102

Routine hematological parameters of the patient over the course of hospital stay		
	21/8/20	3/10/20
Hemoglobin	9.8	11
TLC	4500	8000
DLC	80/18/02	74/24/2
Platelets	2.1 lakh	3.4 lac
Urea	38	32
Creatinine	1.8	0.8
Sodium	140	135
Potassium	3.5	3.9
Calcium	9	9.6
Phosphorus	4.1	3.46
Total bilirubin	0.5	0.13
SGOT	32	29
SGPT	12	29
ALP	113	116
Total protein	4.3	4.8
Albumin	1.03	1.7
Urine R/M	3+ Protein	3+ Protein
24-h urine protein	11 gm	3 gm
Triglyceride	460	392
Total cholesterol	350	287
LDL	230	161
VLDL	82	78
HDL	48	49

The provisional renal biopsy report was suggestive of membranous glomerulonephritis. To rule out secondary membranous nephropathy, autoimmune markers (Antinuclear antibodies [immunofluorescence] (ANA [IF]), Extractable Nuclear Antigen Antibodies (ENA), Antineutrophil cytoplasmic antibodies (ANCA), Rheumatoid factor (RF), anti-Cyclic



Figure 1: Milky chyle drained by thoracentesis

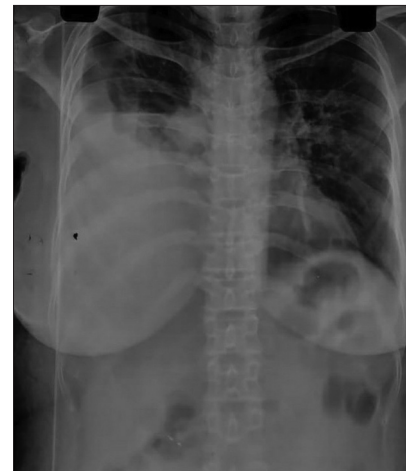


Figure 2: Chest X ray showing right sided pleural effusion

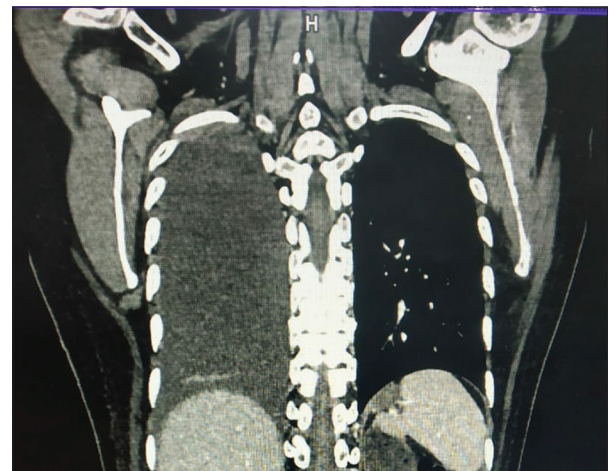


Figure 3: Contrast enhanced computed tomography scan showing right sided pleural effusion

citruinated peptide (CCP) anti-Thyroid peroxidase (TPO) antibody) were sent which were found negative. For infections, serology for HIV, hepatitis B and C, filaria, and syphilis were negative. There was no history of Nonsteroidal anti-

inflammatory drugs (NSAID) abuse and CECT of the chest and abdomen revealed right-sided pleural effusion with moderate ascites and no lymphadenopathy and abdominal or thoracic mass. It revealed left renal vein thrombosis.

The final renal biopsy was suggestive of membranous glomerulonephritis with granular deposits of Immunoglobulin G (IgG) (4+), C3 (3+), C1q (2+). Immunoglobulin M (IgM) and Immunoglobulin A (IgA) were negative and on Immunohistochemistry (IHC), Anti-phospholipase A2 receptor (PLA2R) antibodies (PLA2R) was positive. Serum PLA2R antibody was negative. Finally, diagnosis of primary membranous nephropathy was made with chylous ascites and right-sided chylothorax with left renal vein thrombosis.

She was managed with modified Ponticelli regimen and anticoagulation. After 2 weeks of hospitalization, the patient improved and was discharged with Kidney function tests (KFT): 32/0.8, total protein/albumin: 4.8/1.7, 24 h urine protein: 3 gm international normalized ratio (INR): 2.1.

Discussion

Chylothorax is the occurrence of chyle in the pleural space. The diagnosis is made by pleural fluid analysis which contains a high level of triglyceride (>110 mg/dL) and is confirmed by finding chylomicrons.^[4]

Chylothorax has a variety of causes like trauma: post-surgical or accidental injury. In the absence of trauma, malignancy may account for three-fourths of all cases, lymphoma more commonly than carcinoma.^[5]

Despite extensive evaluation etiology of transudative chylothorax may be unclear.^[6]

Chylous ascites may cause chylothorax in nephrotic syndrome by unidirectional migration of fluid from peritoneal to pleural cavity because of negative intrathoracic pressure during inspiration,^[7] which was also the case in our patient. Although nephrotic syndrome is common in primary care practice, chylothorax secondary to it, is not so common, and therefore, this case report may sensitize the primary care physician about this uncommon manifestation of this relatively common disease.

Lindenbaum and Scheidt reported that chylous ascites is frequently found in patients with nephrotic syndrome. They review 140 cases (97 children and 43 adults) of nephrotic syndrome of various etiologies. They found ascites was present in 90 cases (64%) and paracentesis was done in 35 cases of which 16 (52%) were having chylous milky white ascitic fluid.^[8]

Another mechanism for chylothorax in nephrotic syndrome can be hypercoagulable state causing venous thrombosis (superior vena cava and subclavian vein thrombosis). Rathore V from

India has reported a case of chylothorax in an 8-year-old child secondary to superior vena cava thrombosis.^[9] In our case, there was left renal vein thrombosis but no thrombus was seen in other veins on Computed tomography (CT) angiography.

Before the final biopsy report came, all secondary causes of nephrotic syndrome were ruled out. The lymphangiography of our patient was normal.

The treatment of chylothorax includes treatment of the underlying condition, conservative, and surgical management. In our case, it was nephrotic syndrome causing chylothorax, which was managed with therapeutic thoracentesis for symptomatic relief succeeded by the modified Ponticelli regimen with which our patient improved.

Conclusion

Chylothorax associated with nephrotic syndrome is a very rare disorder, especially in adults. When a patient has chylothorax (especially right-sided) with chylous ascites—nephrotic syndrome should be included in the differential diagnosis. Treating the underlying disease is most important in the management of these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms in which the patient has given her consent for images and other clinical information to be reported in journal. The patient understands that her name and initials will not be published and due effort will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil

Conflicts of interest

There are no conflicts of interest.

References

1. Ayalon R, Beck LH Jr. Membranous nephropathy: Not just a disease for adults. *Pediatr Nephrol* 2015;30:31-9.
2. Moss R, Hinds S, Fedullo AJ. Chylothorax. A complication of the nephrotic syndrome. *Am Rev Respir Dis* 1989;140:1436-7.
3. Chen YC, Kuno MC, Chen HC, Liu MQ, Hwang SJ. Chylous ascites and chylothorax due to the existence of transdiaphragmatic shunting in an adult with nephrotic syndrome. *Nephrol Dial Transplant* 2005;20:1501-2.
4. Hillerdal G. Chylothorax and pseudochylothorax. *Eur Respir J* 1997;10:1157-62.
5. Merrigan BA, Winter DC, O'Sullivan GC. Chylothorax. *Br J Surg* 1997;84:15-20.
6. Rajdev K, Avula A, Sharma D, Mansour W, Agarwal S,

- Siddiqui AH, *et al.* A case of transudative chylothorax: A diagnostic dilemma. *Cureus* 2018;10:e2247.
7. Lieberman FL, Hidemura R, Peters RL, Reynolds TB. Pathogenesis and treatment of hydrothorax complicating cirrhosis with ascites. *Ann Intern Med* 1966;64:341-51.
8. Lindenbaum J, Scheidt SS. Chylous ascites and the nephrotic syndrome. *Am J Med* 1968;44:830-6.
9. Rathore V, Bhattacharya D, Pandey J, Bhatia A, Dawman L, Tiewsoh K. Chylothorax in a child with nephrotic syndrome. *Indian J Nephrol* 2020;30:32-4.