Chylothorax in a Child with Nephrotic Syndrome

Abstract

Chylothorax is an uncommon presentation of venous thrombosis in nephrotic syndrome. We present a case of an 8-year-old boy with nephrotic syndrome who presented with prolonged respiratory difficulty and dry cough. A detailed evaluation revealed left chylothorax secondary to thrombosis of the left brachiocephalic vein. He improved with conservative management including anticoagulation therapy, intercostal chest tube drainage, and dietary modification. This case highlights the need to consider venous thrombosis as a cause of chylothorax in patients with nephrotic syndrome to institute appropriate treatment.

Keywords: *Chylothorax, nephrotic syndrome, thrombosis*

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Introduction

Chylothorax is an uncommon cause of pleural effusion in children. Chyle is a milky fluid containing fat droplets, which drain from lacteals of small intestine into the lymphatic system during digestion. Chylothorax results from leakage of chyle from the lymphatic system into the pleural space. The most common causes in children are thoracic duct injuries (recent surgery, trauma), congenital abnormalities of lymphatics, and malignancies.[1] The less common causes include thrombosis of superior vena cava or subclavian vein. While chylothorax occurring as a result of transdiaphragmatic movement of chylous ascites is well known in patients with nephrotic syndrome,[2-4] chylothorax without ascites due to superior vena cava thrombosis have been infrequently reported in literature.[5] We report a case of a boy with nephrotic syndrome who developed chylothorax secondary to venous thrombosis.

Case Report

An 8-year-old boy was diagnosed with nephrotic syndrome at the age of 3 years. He had infrequent relapses, which were treated with oral prednisolone. He presented to us, after being in remission for the past 3 years, with progressive difficulty in breathing and cough for 3 weeks. On examination, he was tachypneic with respiratory rates of 38/min. His peripheral capillary oxygen saturation was 89%, which improved to 98% with supplemental oxygen. He was afebrile and had normal blood pressure. He had mild facial puffiness but no pedal edema. He had prominent veins on the left infraclavicular aspect of the chest [Figure 1]. His trachea was shifted to the right, with reduced movements of left hemithorax. There was decreased air entry on the left hemithorax with stony dullness on percussion, along with reduced vocal fremitus and resonance. Other systemic examination was unremarkable.

A chest X-ray revealed homogeneous opacity of the left hemithorax with shift of the mediastinum to the right side and obliteration of the left cardiophrenic and costophrenic angles, suggestive of a massive left pleural effusion [Figure 2a]. Diagnostic pleural tap revealed milky fluid [Figure 2b]. Pleural fluid (PF) analysis showed 1260 cells/mm^3 (predominantly lymphocytes, no malignant cells), protein of 239 mg/dL, and sugar of 99 mg/dL. Gram and Ziehl–Neelsen stain were negative, and culture was sterile. PF triglyceride was 337.6 mg/dL and cholesterol was 15.9 mg/dL. PF: serum triglyceride ratio was 1.58, and PF: serum cholesterol ratio was 0.03. Although the gold standard test to prove chyle is lipoprotein analysis demonstrating chylomicrons, yet wherever unavailable, a PF triglyceride >110 mg/dL with a cholesterol of <200 mg/dL is diagnostic.

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Furthermore, milky color of the fluid, being sterile, absolute count of >1000 cells/m^3 (80% lymphocytes), PF: serum triglyceride >1, and PF: serum cholesterol <1 all suggest a diagnosis of chylothorax.[6]

Investigations showed hemoglobin of 13.7 g/dL, total leucocyte count of 8900 cells/mm^3 (neutrophils 72%, lymphocytes 26%, monocytes 2%), platelet count of 320,000/mm^3, serum urea 25 mg/dL, serum creatinine 0.37 mg/dL, serum total proteins 4.8 g/dL, serum albumin 2.7 g/dL, serum cholesterol 467 mg/dL, and serum triglycerides 213 g/dL. Urinalysis revealed 4+ proteinuria and his urine protein excretion was 49.2 mg/m^2/h. Serology for hepatitis B surface antigen, hepatitis C virus, and human immunodeficiency virus was negative. Complement levels (C3 and C4) were normal, and antinuclear antibody was negative. His prothrombin time was 13 s (normal 12–14 s), prothrombin index 100%, international normalized ratio 1.1, and activated partial thromboplastin time was 31 s (normal 25–32 s).

Abdominal ultrasound examination showed normal sized kidney and did not reveal any free fluid in the peritoneum. Contrast-enhanced computerized tomography scan of the chest showed gross left pleural effusion with an underlying passive collapse of left lung, and nonopacified left brachiocephalic vein with a partial filling defect of left superior vena cava suggestive of thrombosis [Figure 3]. Lymphoscintigraphy showed mild inhomogeneous tracer activity in the left supraclavicular region localized at the junction of the left internal jugular and left subclavian vein with nontracer avid massive left pleural effusion.

Chylothorax was managed by intercostal chest tube drainage and a high-calorie (120–140 calories/kg/day), high-protein (2.5–3 g/kg/day), and a low-fat diet rich in medium chain triglycerides (MCTs), with supplements MCT oil. He was started on subcutaneous enoxaparin at a dose of 1 mg/kg/dose twice daily, and the dose was titrated to maintain an anti-factor Xa level of 0.5–1.0 U/mL. Initially, his chest drain output was 2 L/day, which decreased over the next 8 days and chest drain was removed on day 11 of hospital stay.

His relapse of nephrotic syndrome was managed with oral prednisolone (60 mg/m^2/day) for 2 weeks following which he achieved remission. Prednisolone was prescribed at a dose of 40 mg/m^2/every other day for an additional 4 weeks.

At 3 months of follow-up, he is asymptomatic and continues to be in remission after stopping steroids. His chest X-ray showed resolution of chylothorax [Figure 2c]. Subsequent workup for prothrombotic state including protein C, protein S, DNA testing for factor V Leiden mutation, antithrombin functional activity, lupus anticoagulant, anticardiolipin immunoglobulin (Ig) G, and anticardiolipin IgM was negative.

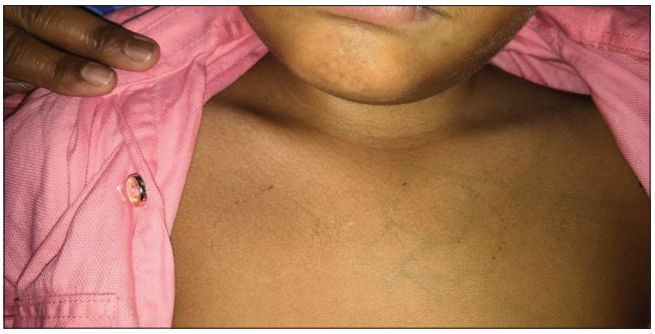

Figure 1: Color photograph showing prominent veins on the left infraclavicular aspect of the chest of the child

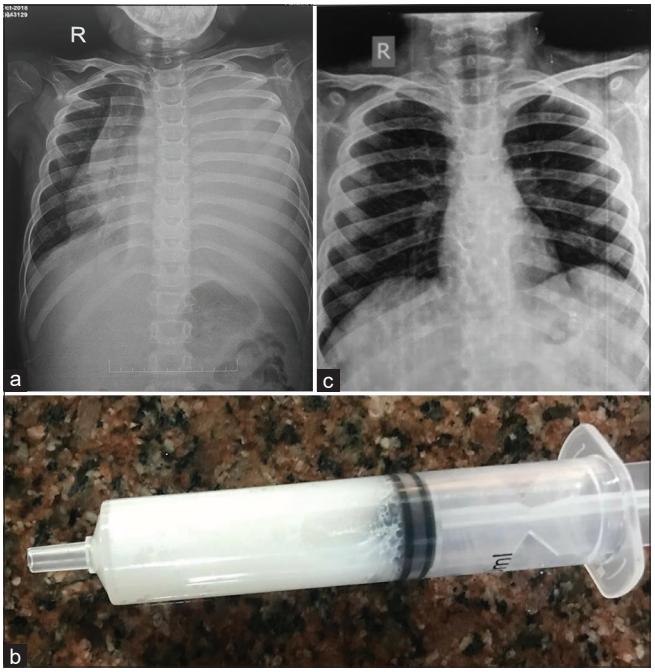

Figure 2: (a) Chest X-ray showing left pleural effusion with mediastinal shift. (b) Color photograph showing milky chyle drained by pleural tap. (c) Chest X-rays showing gradual resolution of effusion on follow-up

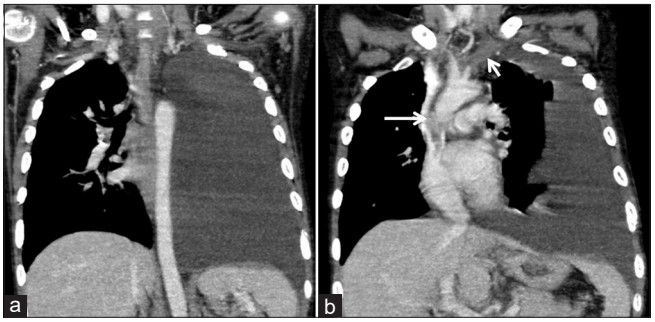

Figure 3: Coronal CECT chest images (a and b) showing gross left-sided pleural effusion. Hypodense filling defect (long arrow in b) suggestive of thrombus is seen in the superior vena cava along its medial wall in its entire course. Superiorly, there is complete thrombotic occlusion of the visualized left brachiocephalic vein (short arrow in b)

Discussion

Chylothorax in children is usually due to trauma following thoracic surgeries. Thrombosis of central venous system

leading to chylothorax is rare and is typically secondary to central venous catheterization.[7] Nephrotic syndrome is a well-known hypercoagulable state owing to loss of anticoagulant proteins in the urine and has been associated with a variety of thromboembolic events including renal vein thrombosis, pulmonary embolus, sagittal sinus thrombosis, and thrombosis of the indwelling arterial and venous catheter.[8,9] However, thrombosis of central veins leading to chylothorax in nephrotic syndrome has been infrequently reported.[5,10,11] One of the clinical clues which led us to suspect the possibility of central vein obstruction in our child was the presence of dilated veins in the thorax, highlighting the importance of astute clinical examination.

Management of chylothorax usually requires intercostal chest tube drainage to relieve respiratory symptoms. Intercostal chest tube drainage also helps in recording the rate of chyle leak and aids in assessment of response to therapy. Medical management consists of providing a low-fat, high-protein, and a high-calorie diet. This diet not only replaces fluid, electrolytes, and protein lost in chyle and prevents malnutrition but also decreases the lymph leak. Also, medium-chain triglycerides (MCTs) are absorbed directly into the portal system rather than the intestinal lymphatics, thereby are invaluable in providing adequate calories. Our child had responded to the above measures. Failure to respond to these measures may require the use of somatostatin analog, total parenteral nutrition, or surgical correction.[12]

Treatment of the underlying cause of chylothorax is essential. Venous thrombosis is usually treated with anticoagulation with unfractionated heparin, low-molecular-weight heparin, or oral anticoagulants. Our child was treated with enoxaparin for 3 months with a target to maintain an anti-factor Xa level of 0.5–1.0 U/mL. Anticoagulation was stopped after 3 months as the child had entered remission with steroids and tests for the congenital prothrombotic state were negative.[13]

To conclude, chylothorax in a child with nephrotic syndrome is a rare presentation of thoracic vein thrombosis. A high index of suspicion and appropriate workup is needed to reveal the etiology of chylothorax.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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