STUDENT CASE REPORT

A Rare Case of Persistent Chylothorax

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ABSTRACT

Chylothorax is a condition referring to the presence of chyle in one or both pleural cavities. Chylothorax is an especially rare condition in the pediatric population. It most often occurs secondary to infection, iatrogenic or non-iatrogenic trauma, malignancy, or congenital malformations. Prompt diagnosis and effective treatment of chylothorax in a pediatric patient is essential as this condition is associated with significant morbidity and mortality. This case describes a persistent chylothorax in a 12-year-old boy who initially presented to a community hospital emergency room complaining of chest pain. Radiologic examinations included chest X-rays, ultrasounds, and computed tomography scans. After failed conservative management, the patient was eventually transferred to a tertiary academic institution for more specialized care. Additional testing was performed, including lymphoscintigraphy and genetic panels. Results of lymphoscintigraphy were normal, while genetic testing showed four variants of undetermined significance. We discuss the ongoing medical management and maintenance of a pediatric patient who remains stable with persistent chylothorax.

CASE REPORT

We present a case of chylothorax in a previously healthy 12-year-old boy who initially presented to his primary care provider (PCP) with chest pain. The patient reported intermittent chest pain for approximately one month. He was diagnosed with pectus excavatum and referred to pediatric cardiology for further evaluation. An echocardiogram performed by the cardiologist revealed an incidental right pleural effusion, which prompted the patient's immediate referral to the community hospital emergency department.

Upon arrival, a thorough history was obtained. The patient's mother reported that his symptoms may be related to a fall from his bicycle approximately 3-4 months prior, in which the patient flew over the handlebars and landed on his chest. The intermittent chest pain had no exacerbating or alleviating factors. He denied any additional symptoms, including shortness of breath, difficulty breathing, palpitations, recent or current fever, chest pain with exertion, syncope, night sweats, weight loss, or cough. Physical exam was unremarkable besides a pectus excavatum deformity. Pulmonary auscultation was significant for an expiratory wheeze in the

posterior right upper lung. Vital signs were normal for age. Laboratory studies were within normal limits except for mild anemia (hemoglobin 12.8 g/dL, hematocrit 38.7%) and an elevated alkaline phosphatase level (561 U/L). An electrocardiogram (ECG) on initial evaluation revealed a right bundle branch block.

Several imaging modalities were utilized, including chest X-ray (CXR), ultrasound (US) pleural effusion survey, and computed tomography (CT) of the thorax, abdomen, and pelvis. CXR revealed a patchy atelectasis or infiltrate in the right middle lobe **(Figure 1)**. CT revealed an airspace opacity in the right perihilar region and pleural effusion on the right. **(Figure 2).** US confirmed these findings, indicating a right pleural effusion with a volume of approximately 900 mL. The patient was admitted to the pediatric hospitalist service for continued management. Thoracentesis was performed with chest tube placement, producing 550 mL of milky fluid. Initial conservative management included a low-fat diet, and the chest tube was set to suction.

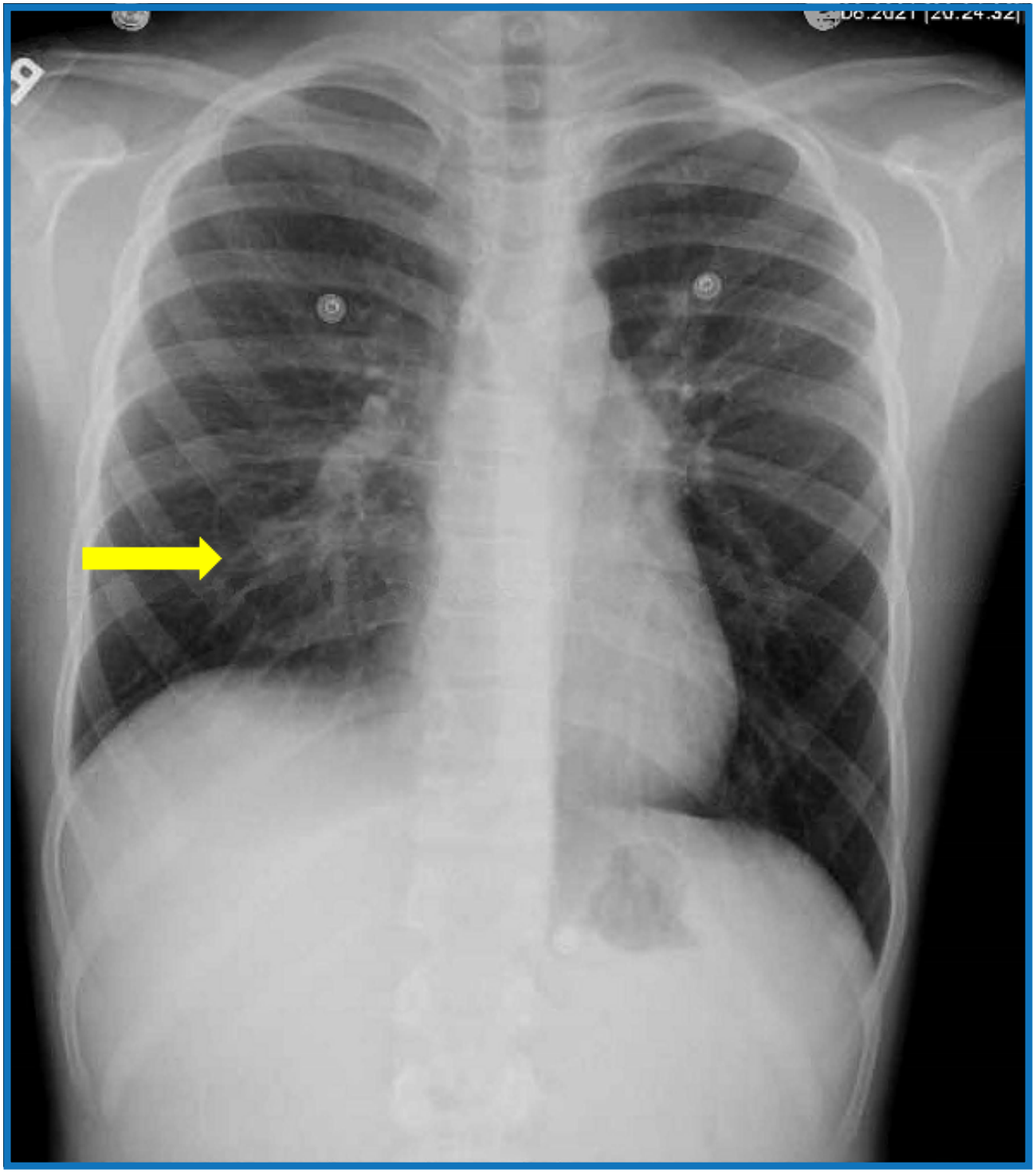

Figure 1: Chest X-ray showing a questionable pectus excavatum deformity, and patchy atelectasis or infiltrate of the right middle lobe.

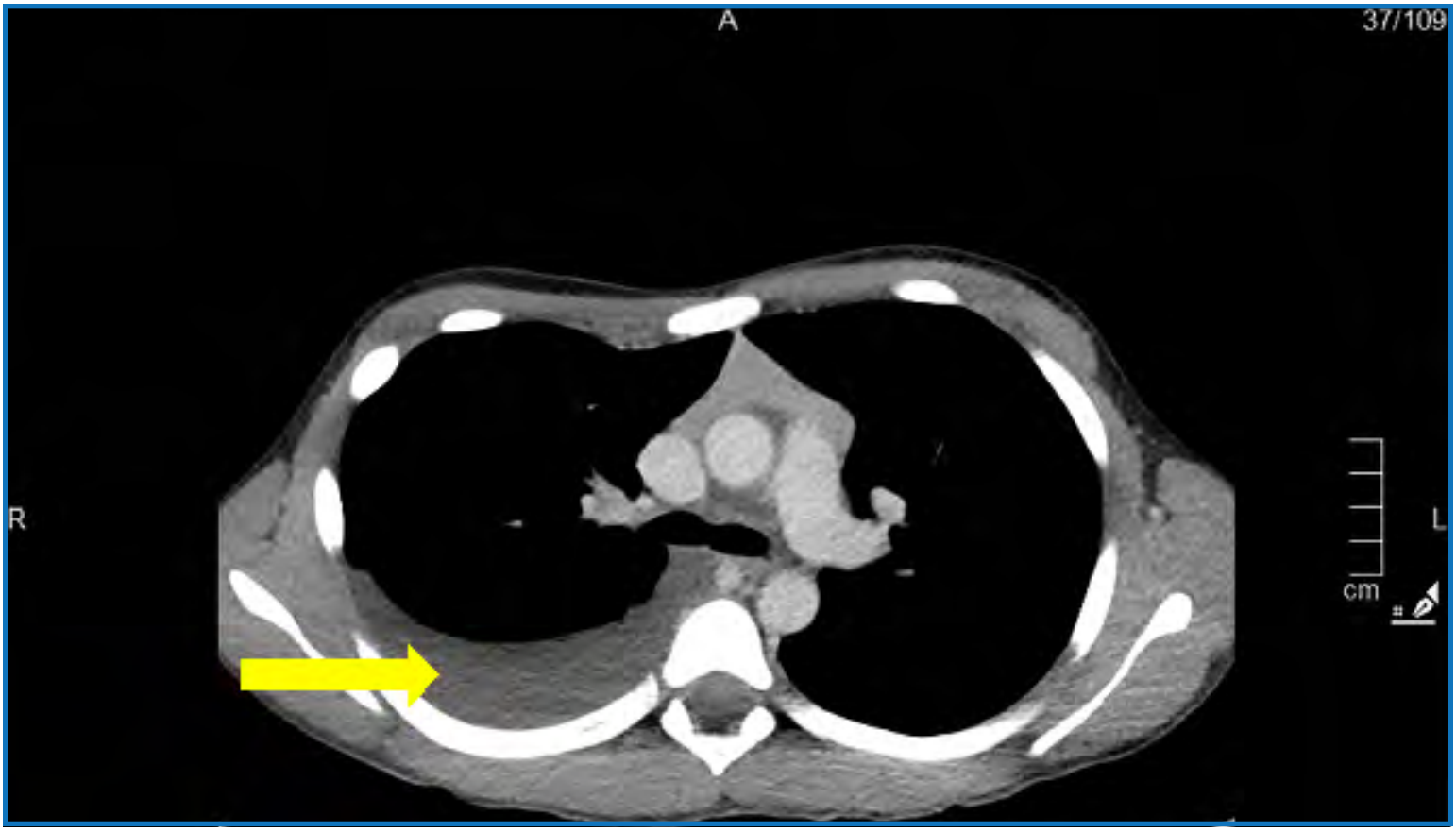

Figure 2: CT imaging significant for right-sided pleural effusion.

Several pleural fluid analyses confirmed the presence of a chylothorax. Results showed amylase 12 U/L, glucose 155 mg/dL, lactate dehydrogenase (LDH) 144 U/L, and triglycerides 1887 mg/dL. The pleural fluid triglyceride concentration above 110 mg/dL was sufficient to confirm the fluid as chylous.[4] Additionally, chyle is a lymphocyte-dominant fluid with cell counts of 80% lymphocytes or higher.[4] The pleural fluid cytology in this case was significant for a lymphocyte count of 99%, further confirming a chylothorax. Flow cytometry of pleural fluid showed no immunophenotypic abnormalities, and cultures for any infectious etiology were negative. Additional hematologic analysis showed normal immunoglobulin levels, LDH, uric acid, C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR). Flow cytometry of peripheral blood indicated no presence of a non-Hodgkin's lymphoma or acute leukemia. QuantiFERON and heterophile antibody testing proved negative for tuberculosis and infectious mononucleosis, respectively. These analyses presumably ruled out infectious and malignant causes of chylothorax.

Conservative management was continued for the subsequent 1-week period that the patient remained admitted in the community setting. A total of 720 mL of chylous fluid drained from the chest tube over the next five days. Due to the failure of chest tube output to cease, the patient was transferred to a tertiary academic pediatric center for subspecialty care.

Upon arrival, a peripherally inserted central catheter (PICC) line was placed to deliver total parenteral nutrition (TPN) with intralipids and continuous intravenous Octreotide. The patient was only allowed oral medium chain triglyceride (MCT) oil and clear liquids. Despite these interventions, the chylothorax persisted. Lymphoscintigraphy was performed, which showed no evidence of extravasation. Repeat CT imaging confirmed the presence of a persistent, low-output right pleural effusion. The treatment plan consisted of IV octreotide, TPN and lipids, and chest tube output monitoring, which was continued for 14 days after transfer.

Despite the persistence of the chylothorax, daily chest tube output gradually subsided over the 14 days. This gradual improvement prompted the discontinuation of TPN and IV octreotide. During this time, a consultation was performed by the clinical geneticist. Physical exam features, including myopia, high-arched palate, positive Marfan (Walker–Murdoch) wrist sign, pectus excavatum, and a borderline large aorta seen on cardiac echo, prompted ordering a DNA genetics panel, which would investigate for possible Marfan syndrome, Noonan Syndrome, and other connective tissue disorders.

The patient was discharged 19 days after transfer. Upon follow-up with pulmonology one month later, the patient was readmitted to the hospital after imaging revealed a persistent right-sided pleural effusion. The patient was asymptomatic and cleared for discharge. Follow-up after one month showed a stable chylothorax.

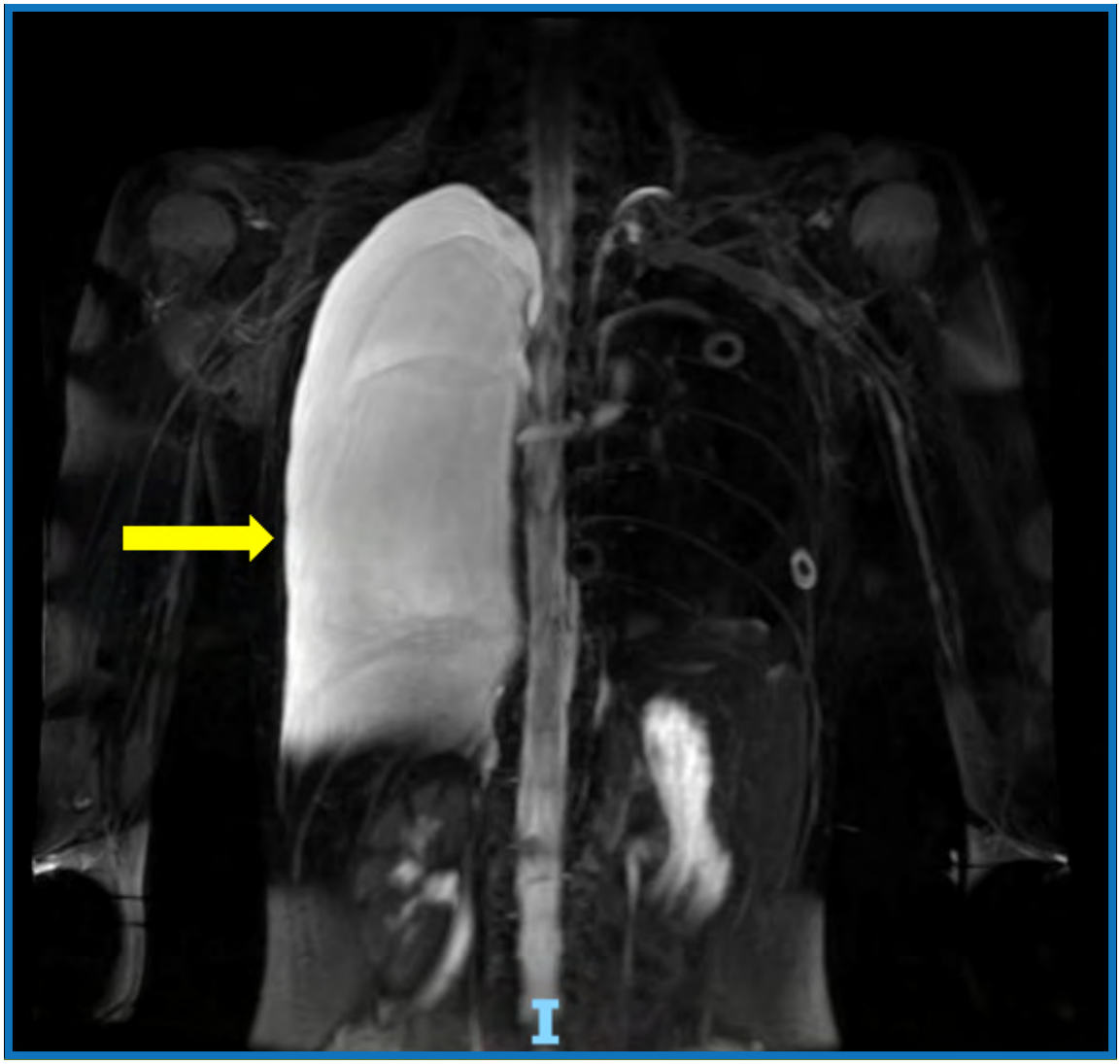

Figure 3: MR lymphangiogram demonstrating a large right pleural effusion

Genetic testing revealed four variants of undetermined significance in the following genes: ABL1, ADAMTS17, B4GALT7, and C1S. The variant in one copy of the ABL1 gene may cause symptoms as it follows an autosomal dominant pattern. This gene is associated with various skeletal malformations and congenital heart defects. The single variants of undetermined significance in the subsequent genes are all inherited in a recessive manner and, as such, are unlikely to contribute to the patient's features. Further genetic counseling was recommended, as well as surgical correction of the patient's pectus deformity and a possible thoracic duct embolization if complete resolution of chylothorax cannot be achieved in the future.

In the months following, the patient underwent MR and intranodal lymphangiograms, which showed no evidence of lymphatic leakage. The studies demonstrated good flow from the thoracic duct into the left subclavian vein without any evidence of obstruction. He continues to require bimonthly thoracentesis. It was determined by the interventional radiology specialists that he likely does have a leak in his lymphatic system but that it is too small to be visualized. While the presentation was initially concerning for an underlying complicated vascular anomaly, none was found. Ultimately, the lymphatic leak was attributed to trauma from his bicycle accident as the most likely etiology.

DISCUSSION

Chylothorax refers to the accumulation of chylous fluid in the pleural spaces. This condition is exceptionally rare in children and adolescents and has the potential to cause significant morbidity and mortality. Chylothorax can lead to significant respiratory distress, fluid imbalances, malnutrition, and secondary immunodeficiency if not appropriately diagnosed and treated.[2,5] Although chylothorax is a rare and challenging condition to identify, its diagnosis is crucial as untreated cases are almost always fatal.[6]

The diagnosis of chylothorax is facilitated initially through imaging modalities, including X-ray, ultrasound, and CT scan, which are used to identify fluid collections and assess the size and location of pleural effusion. Once identified, chyle obtained via thoracentesis is typically characterized by a white, odorless, milky pleural fluid. The specific composition of the fluid is characterized by a high

triglyceride content, elevated protein, and albumin level, along with a high cell count (>1,000 cells/mcL) predominately composed of >80% lymphocytes.[7]

There is a diverse array of etiologies to consider in diagnosing a pediatric chylothorax. Congenital etiologies include lymphatic malformations and genetic syndromes such as Noonan Syndrome, Turner Syndrome, and Gorham-Stout Syndrome. Traumatic chylothoraces often result from injury during operations for congenital heart malformations, lymph node excisions, and therapeutic procedures such as subclavian vein catheterization. Other traumatic causes include blunt or penetrating injuries and even actions like coughing and vomiting that may induce a chylothorax. Additionally, malignant etiologies must be considered, including conditions such as neurogenic tumors and lymphomas. Finally, granulomatous infections such as tuberculosis must be thought of as well.[2,8]

Specifically, in this case, with the exclusion of malignant and infectious etiologies through laboratory testing, we were left to consider a traumatic fall from a bicycle 3-4 months before the patient's onset of chest pain. Of note, literature shows that a traumatic chylothorax usually presents after a latent period of 2 to 10 days between the trauma and the onset of pleural effusion.[2,9] With normal lymphoscintigraphy, lymphatic malformation was also largely ruled out.

As in this case, initial management of chylothorax in a pediatric patient should begin conservatively to relieve respiratory symptoms, prevent recurrence, and treat or avoid malnutrition and immunodeficiency.[6] Treatment should begin with chest tube placement and initiation of a low-fat diet with MCT oil supplementation to reduce lymphatic production.[7,10] TPN with IV intralipids should be started if the chyle output does not subside. Additionally, a trial of octreotide or somatostatin can be started as these medications have been shown to reduce lymphatic fluid production via vasoconstriction of the splanchnic circulation.[2] If chylothorax persists after 2-4 weeks of conservative management, surgical management with thoracic duct ligation, pleurodesis, or pleuroperitoneal shunt placement should be considered.[2,5] Of note, a lymphatic leak will sometimes close after a lymphangiogram with ethiodol by causing an embolic effect at the leak site.

CONCLUSION

Chylothorax is a rare and life-threatening condition in the pediatric population. With a wide array of etiologies, clinicians must thoroughly explore many potential congenital, traumatic, malignant, and infectious causes. Identification and treatment of this condition must be prompt and effective, as chylothorax is associated with high morbidity and mortality. This case highlights a standard procedure for managing persistent chylothorax in a child.

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