

Case Report**A LARGE RECURRENT RETROPERITONEAL LYMPHANGIOMA IN A PREVIOUSLY HEALTHY ADOLESCENT MALE**Samer Bou Karroum^{1*}, Aya Bou Fakhreddine², Raphael Mattamal¹**Author information:** ¹Departement of Pediatrics, Texas Tech University Health Sciences Center, Amarillo, Texas, ²Center of Research Institute, Texas Tech University Health Sciences Center, Lubbock, Texas

Received: 10-29-2023; Accepted: 12-05-2023; Published: 12-15-2023.

Abstract: Lymphangiomas are uncommon benign lymphatic vessel tumors that can affect any area of the body served with the lymphatic system. Intra-abdominal lymphangiomas are considered rare, accounting for less than 5 percent of all lymphangioma cases. In this case report, we present a recurrent large retroperitoneal lymphangioma in a previously healthy 15-year-old boy. The patient presented to the Emergency Department with abdominal pain lasting for one week. The pain, described as diffuse, crampy, and non-radiating, gradually increased with time, and was aggravated by movement. A large retroperitoneal fluid collection was found on the Computed Tomography (CT) scan. Subsequently, the patient underwent CT-guided drainage, at which 700ml of bloody fluid was extracted. After three months, the patient was readmitted to the hospital for a similar presentation of abdominal fullness and pain. Complete blood count analysis indicated an unremarkable profile, except for a left shift of 82.2% neutrophils and a white blood cell count of 11,300/mcl. A CT scan of the abdomen reported a large, thin-walled cystic lesion in the right hemi-abdomen with minimal thin internal septations, resulting in moderate right hydronephrosis/hydroureter.

Keywords: Retroperitoneal Lymphangioma, Adolescent boy, Abdominal Pain

INTRODUCTION Lymphangiomas are uncommon benign tumors affecting the lymphatic vessels. While the exact etiology is not fully understood, these tumors are recognized as primary malformations of the lymphatics involving obstructed or insufficient efferent channels [1]. Lymphangiomas can affect any area of the body served by the lymphatic system, with a predominant occurrence (75%) in the neck, head, and axilla [1]. Intra-abdominal lymphangiomas, although rare, account for less than 5 percent of all lymphangioma cases, with a higher incidence in males [1, 2].

Despite their benign nature, lymphangiomas cause symptoms due to mass effects and consequential pressure on surrounding organs. In cases of abdominal lymphangiomas, the space available in the abdominal cavity allows the tumor to grow while the patient remains

asymptomatic for an extended period. The most common locations of abdominal lymphangiomas include the mesentery followed by omentum, mesocolon, and rarely in the retroperitoneum [1, 2].

The etiology of retroperitoneal lymphangiomas suggests a lack of communication between retroperitoneal lymphatic tissue and systemic lymphatic vessels. The clinical presentation of retroperitoneal lymphangioma can range from incidental discoveries to symptoms such as fatigue, fever, abdominal pain, back pain, and hematuria. Imaging modalities, including sonography and computed tomography, can demonstrate lymphangioma; however, a histological exam remains imperative for a definitive diagnosis [3,4]. While most lymphangiomas are evident by age five, many cases do not manifest until adulthood [1].

This case report highlights a rare occurrence of a recurrent large retroperitoneal lymphangioma in a previously healthy 15-year-old male while emphasizing the diagnostic and treatment challenges.

*Corresponding author: Samer Bou Karroum, MD
Departement of Pediatrics, Texas Tech University Health Sciences Center, Amarillo, Texas. E-mail: Samer.Bou-Karroum@ttuhsc.edu

CASE: A 15-year-old male presented to the Emergency Department (ED) with a one-week history of diffuse, crampy, non-radiating abdominal pain. The pain increased with time and was aggravated by movement. The patient reported no bowel or urinary output alterations, frequency, or color. He had no pertinent surgical or medical history and had no other symptoms such as fever, weight loss, appetite change, vomiting, diarrhea, cough, or shortness of breath. He was not on any chronic medications and did not take any medications for abdominal pain. The patient reported occasional nicotine vaping and daily marijuana use over the past year. Moreover, he had no known family history of cancerous abdominal masses. The ED evaluation, including Complete Blood Count and Complete Metabolic Panel, revealed unremarkable results. However, a Computed Tomography (CT) scan showed a large retroperitoneal fluid collection. The patient underwent CT-guided drainage of the fluid, at which 700 ml of bloody fluid was extracted (Picture 1). The drainage provided symptomatic relief, and the patient was discharged.

Three months later, the patient was readmitted to the hospital for a similar presentation of recurring abdominal fullness and pain that has been gradually increasing with time. This pain was accompanied by urinary urgency and decreased appetite. Physical examination revealed a distended abdomen with fullness on palpation, even though no hepatosplenomegaly was reported.

Laboratory findings indicated a left shift of 82.2% neutrophils, a white blood cell count of 11,300/MCL, hemoglobin of 12.4 g/dl, and hematocrit of 36%. The complete metabolic panel showed normal electrolytes with mildly decreased albumin of 3.1 g/dl. Other tests, including lipase level and urinalysis, were unremarkable. Prothrombin time (PT) and Partial Thromboplastin Time (PTT) were also within normal limits. Carbohydrate Antigen 19-9 (CA 19-9) tumor marker (4 units/ml) as well as Carcinoembryonic Antigen (CEA of 0.3 ng/ml) were low. A repeat CT scan of the abdomen reported a large thin-walled cystic lesion in the right hemi-abdomen with minimal thin internal septations, resulting in moderate right hydronephrosis/hydroureter. The differential diagnosis after the abdominal CT scan included peritoneal inclusion cyst or lymphangioma. Further work-up and clinical correlation were recommended.

Abdominal fluid analysis revealed a brown turbid fluid. It included 29,117 cells/mcL White blood cells, 1,601,400

cells/mcL Red Blood Cells, 83 cells/mcl segmented cells, and 12 Lymphocytes. Microbiology cultures, including anaerobic culture, gram stain, and fungal culture with smear, were all negative. Tissue pathology was negative for malignant cells or atypical lymphocytes. The report described hemosiderin macrophages and degenerate blood, a mixed population of small reactive T-cell and B-cell lymphocytes with fragments of benign connective tissue, skeletal muscle, and fat. The beta HCG was less than 1 million international units/ml. Given the benign picture with no evidence of malignancy, as well as the CT scan findings, the patient was diagnosed with cystic lymphangioma of the abdomen.

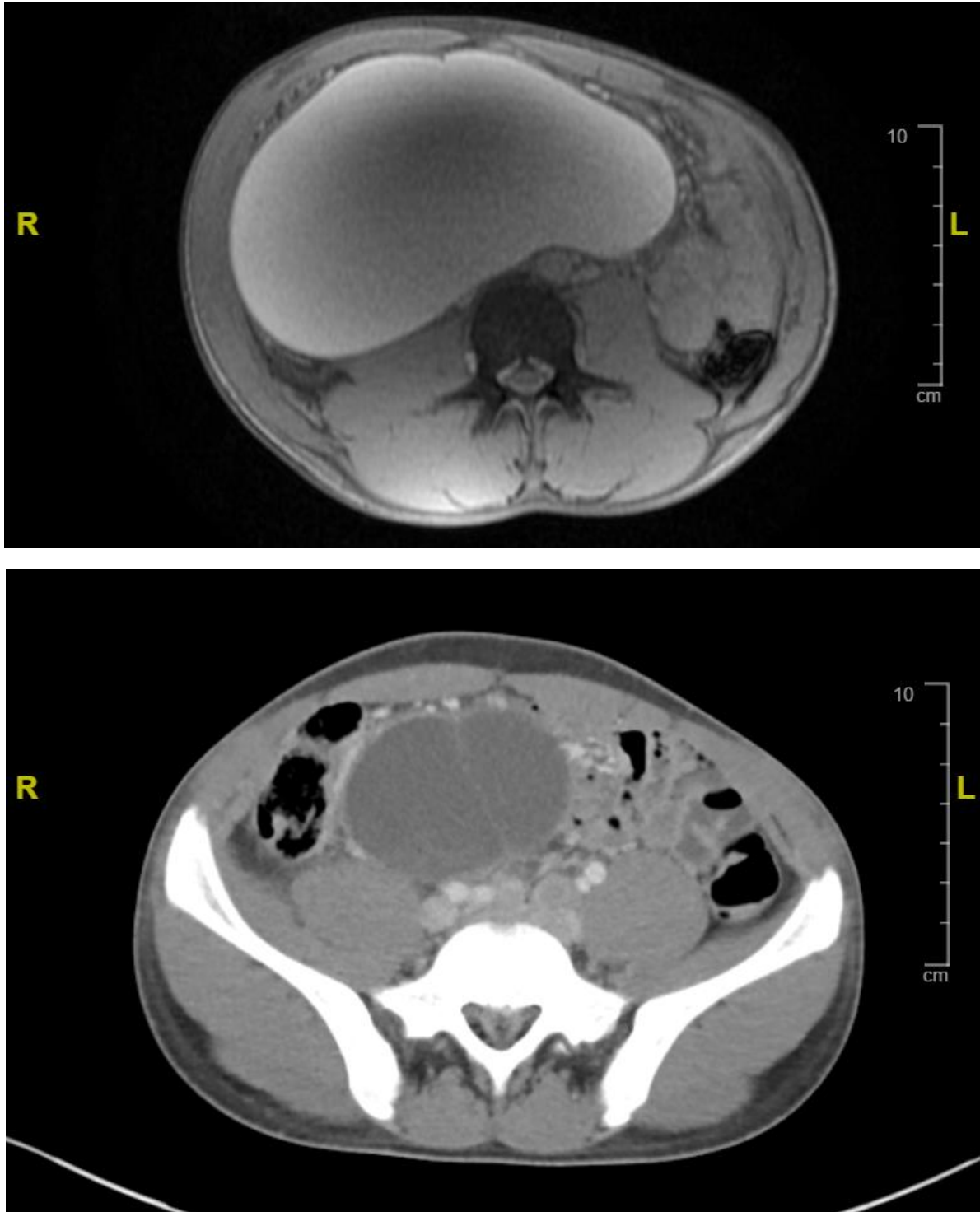
Upon an Interventional Radiology consult, fluid drainage with sclerosing agent injection was recommended. One week later, the patient had an abdominal catheter inserted, and a small volume of residual serous fluid was removed. Next, under intermittent fluoroscopy, a diluted concentration of Isovue-300 contrast and sterile water was injected until the fluid cavity was filled at 200mL. The fluid was drained, and 2 mL of Sotradecol (Sodium tetradecyl sulfate sclerosing agent) was prepared into a foam solution and injected to dwell. In comparison, the doxycycline decision was calculated according to the fluid cavity size at a concentration of 10 mg/mL. 1000 mg of doxycycline was injected into the fluid cavity and left to dwell for 2 hours. The patient was discharged with regular follow-up. One-year post-procedure, the patient remains stable with no recurrent lymphangiomas, underscoring the successful management of this rare case.

DISCUSSION Acute abdominal pain accounts for 9% of pediatric visits to primary care providers [4]. Approximately 10% of children aged 4 to 18 years old experienced at least one episode of acute abdominal pain requiring medical attention. While the most common causes of abdominal pain are due to common etiologies such as constipation, gastroenteritis, and appendicitis [4], clinicians should keep an open mind for other less conventional etiologies, particularly when signs and symptoms of the patient do not fit the regular pattern of these common diseases. Thus, a thorough history and physical examination remain pivotal in guiding physicians toward decision-making and diagnosis. An astute physician should formulate a plan based on the most likely etiology to expedite the diagnostic process.

In rare situations, as observed in the presented case, the etiology is neither common nor straightforward.

Lymphangiomas are rare, congenital malformations of the lymphatic system, manifesting predominantly in childhood. Located in different spaces in the body,

lymphangiomas can also appear sporadically in the retroperitoneal space of the abdomen, causing abdominal symptoms.



Picture 1. Thin-walled cystic lesion in the right hemi-abdomen measuring 18 cm transverse by 10 cm AP by 19 cm craniocaudal with minimal thin internal septations.

Malformations originate from lymphatic cisterns in the deep subcutaneous plane, disconnected from the normal lymphatic network. These cisterns, thought to stem from primitive lymph sacs that failed to integrate during embryonic development, are lined with muscle fibers, causing rhythmic contractions. Lymphangiomas, presenting with equal prevalence across races and genders, are typically benign and often evident by age 5, with 50% detectable at birth. The typical appearance on the CT scan is a large, thin-walled, multiseptated cystic masses [5]. Their prognosis is generally excellent, distinguishing them from other conditions such as cutaneous melanoma or neurofibromatosis. Treatment options include surgical excision, vaporization with carbon dioxide lasers, and potentially off-label use of medications like propranolol and sodium tetradecyl sulfate.

CONCLUSION. A physician needs to keep an open-minded approach and explore uncommon etiologies for abdominal pain, recognizing that an early diagnosis of lymphangiomas, though uncommon, can significantly alleviate pain, stress, and efforts for patients.

REFERENCES

1. Levine C, Primary disorders of the lymphatic vessels--a unified concept. *J Pediatr Surg.* 1989;24(3):233-40. doi: 10.1016/s0022-3468(89)80001-62.
2. Shayesteh S, Salimian KJ, Fouladi DF, Blanco A, Fishman EK, Kawamoto S, Intra-abdominal lymphangioma: A case report. *Radiol Case Rep;*16(1):123-127. doi: 10.1016/j.radcr.2020.10.052.3.
3. Thiam O, Faye PM, Niasse A, et al., Cystic mesenteric lymphangioma: A case report. *Int J Surg Case Rep, Int J Surg Case Rep.* 2019;61:318-321. doi: 10.1016/j.ijscr.2019.07.051.
4. Reust CE, Williams A, Acute Abdominal Pain in Children. *Am Fam Physician.* 2016;15;93(10):830-6.5. *J Pediatr Surg.* 2008;43(6):1206-8. doi: 10.1016/j.jpedsurg.2008.01.075.
5. Weeda VB, Booij KA, Aronson DC, Mesenteric cystic lymphangioma: a congenital and an acquired anomaly? Two cases and a review of the literature. *J Pediatr Surg.* 2008;43(6):1206-8. doi: 10.1016/j.jpedsurg.2008.01.075.