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Inguinal prolapse of a retroperitoneal lymphovascular malformation

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ABSTRACT

Abdominal lymphovascular malformations (ALMs) are rare cystic masses that can present with nonspecific symptoms. We present a case of a 7-month-old boy who, during an uncomplicated communicating hydrocele repair, was found to have an incidental large, prolapsed mesenteric abdominal lymphovascular malformation. The case serves to highlight the variability in presentation and natural history of ALMs, and the ease with which they can be disguised by more common pathology. We further underscore the importance of individualized therapy with regards to ALMs, emphasized by our course of active surveillance allowing our patient to avoid ionizing radiation and additional surgical intervention.

1. Introduction

A 7-month-old Caucasian boy with no pertinent past medical or surgical history presented with intermittent right inguinal and scrotal swelling. His birth history was unremarkable, and he had been meeting all developmental milestones. His parents reported frequent changes in size throughout the day. On exam, he had a large, right-sided, non-tender, fluctuant hydrocele that was reducible. It transilluminated, readily communicated, and after reduction, rapidly refilled. Elective repair as well a diagnostic inguinal laparoscopy to evaluate for a contralateral patent processus vaginalis was recommended.

The operation began uneventfully, with the hernia sac being isolated from the testis and cord without difficulty. During diagnostic laparoscopy, the contralateral internal inguinal ring was noted to be closed. While preparing to ligate the processus vaginalis, an adjacent sac was noted at the internal ring and extending along-side the processus. When opened, it drained clear yellow fluid and was seen to extend into the retroperitoneum. It was definitively differentiated from the bladder. Using a larger scope, we re-examined the peritoneal cavity laparoscopically and a soft catheter placed through the separate structure was seen within the cystic structure behind the peritoneum. (Video) Further inspection demonstrated a loculated, cystic structure posterior to and intertwined with the bowel. The malformation had prolapsed through the inguinal ring and extended cranially to the caudate lobe of the liver.

The operation continued with ligation of the processus vaginalis and malformation at the internal ring with a plan for postoperative imaging to guide further decision making. Specimens were sent to pathology and the rest of the operation concluded without any complications.

The CT findings in Fig. 1 were noted to be most consistent with a mesenteric lymphatic malformation with septal calcifications.

Histological and immunohistochemical features were consistent with a lymphatic malformation with interwoven tissue specific for a hernia sac. The lymphovascular malformation tissue was made up of vascular fibroadipose tissue and smooth muscle lined fragments with fibro-intimal proliferation that stained positive with CD31 and D2-40, which fits the clinical picture of an abdominal lymphovascular malformation. The patient was seen in interventional radiology clinic and sclerotherapy was the recommended; however, due to the age of the patient (<1 year), it was advised delay until at least 1 year of age due to the use of bleomycin in the sclerotherapy treatment.

Follow-up imaging with MRI (Fig. 2) a significant decrease in the size of the mesenteric lymphovascular malformation was noted. The patient was asymptomatic and without complications from his hydrocele repair or the lymphovascular malformation. Given the substantial decrease in size on imaging and lack of any symptoms, watchful waiting was recommended.

2. Discussion

We report an incidentally discovered mesenteric lymphovascular malformation that had prolapsed through the internal inguinal ring alongside an inguinal hernia and hydrocele and extended cephalad to the caudate lobe of the liver in an infant. The malformation has

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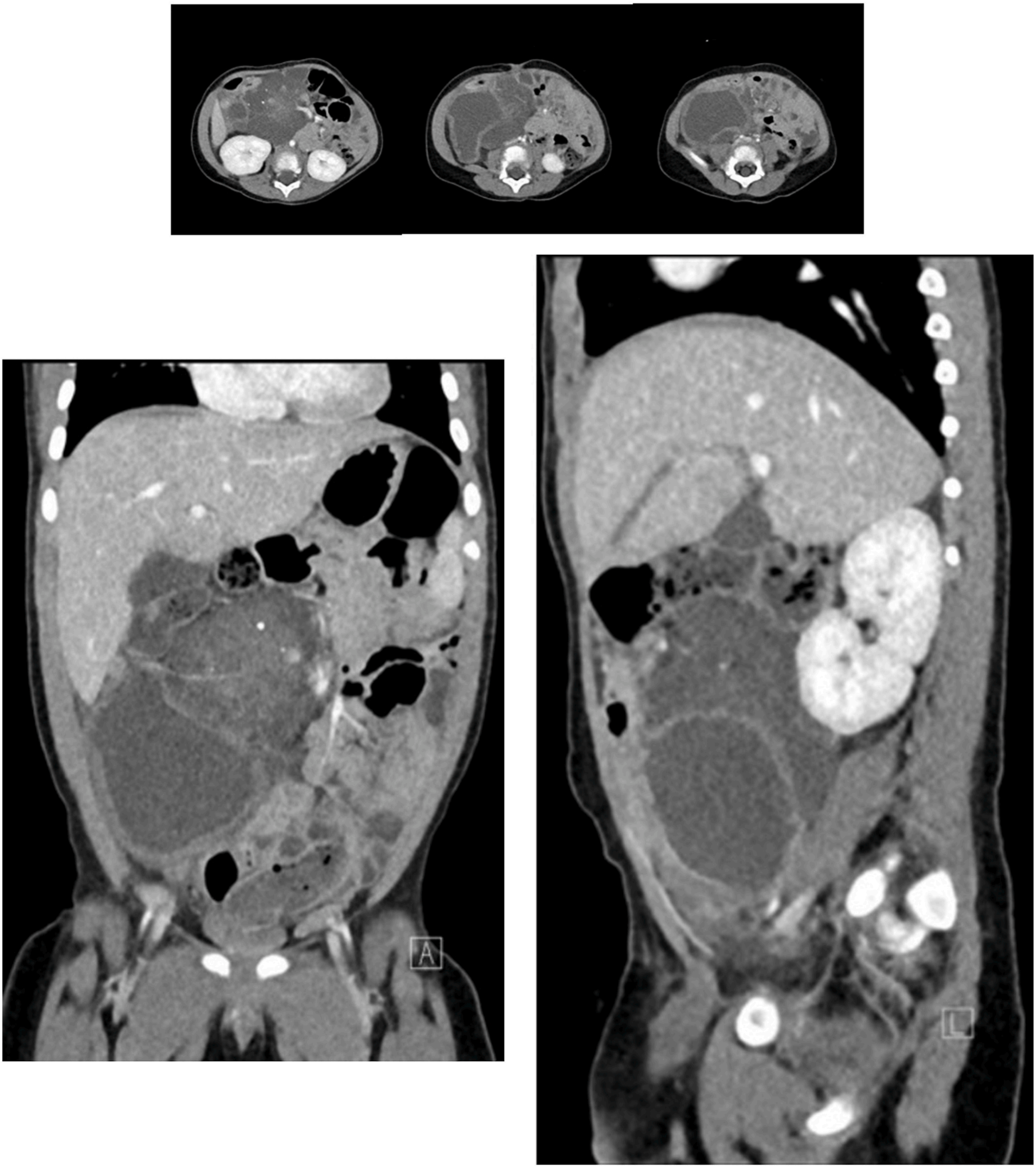

Fig. 1. Post-operative CT imaging demonstrated a 6.3 x 4.8 × 7.1 cm fluid density mass with internal calcifications that encompassed mesenteric vessels and displaced loops of small and large bowels. Additional multiple ovoid enhancing soft tissue structures were identified as well as a thick-walled cystic structure in the right hemiabdomen. A. Axial images B. Coronal image C. Sagittal image.

decreased in size over time without intervention. This case is unique due to the transinguinal prolapse and incidental detection of the abdominal lymphovascular malformation, as well as the age of the patient, location and natural history of the malformation.

Abdominal lymphovascular malformations (ALMs) are thin-walled endothelial channels consisting of lymphatic fluid that can arise associated with low lymphatic flow due to a combination of genetic predisposition and environmental factors.[1,2] These malformations are classified according to the International Society for the Study of Vascular Anomalies based on the presence or absence of cysts, circulation pattern of chyle, and specific characteristics of the lymph itself.[3] The incidence of lymphatic malformations is low, ranging from 1:2,000 to 1:16,000,

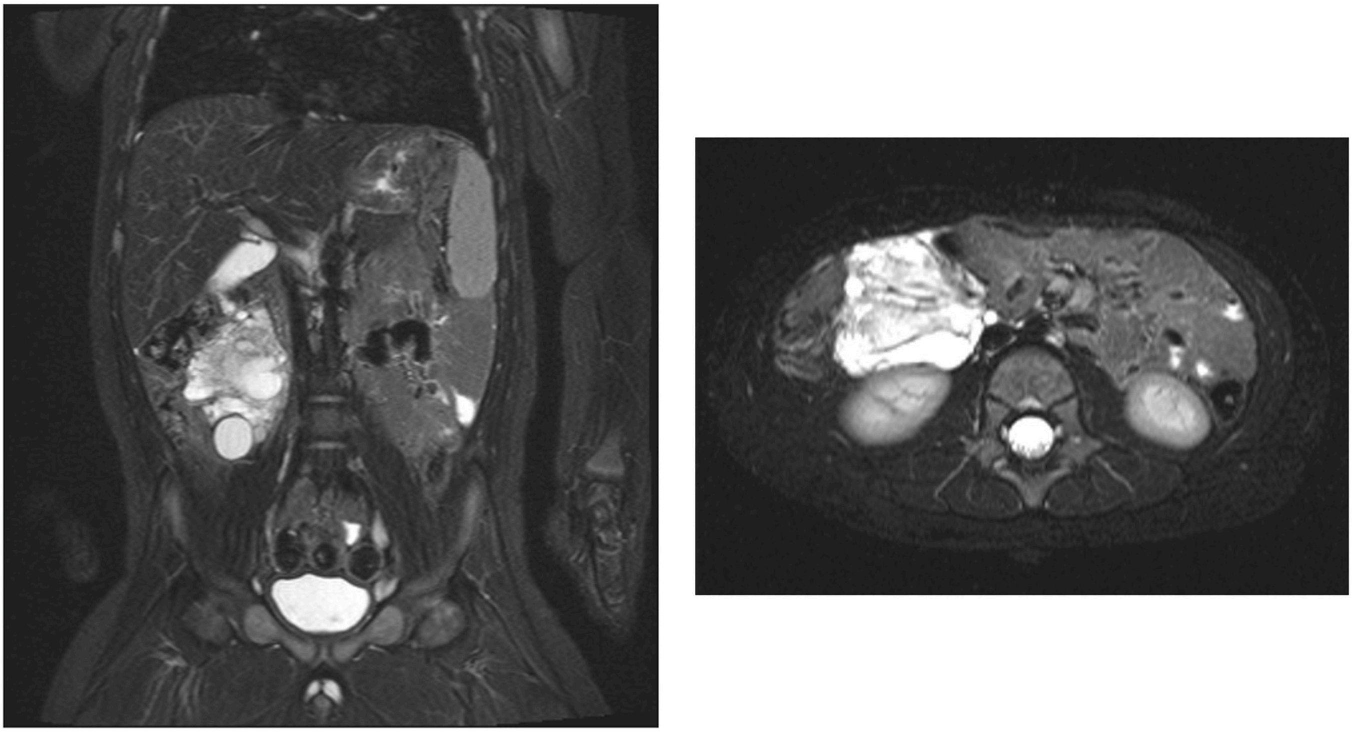

Fig. 2. MR study 4 months post-operatively showing the previously identified veno-lymphatic malformation significantly decreased in size compared to previous CT. The mass has decreased to approximately 3.7 cm AP x 3.9 cm transverse x 6.9 cm craniocaudal. The macrocystic portion of the lesion has also significantly decreased in size by several centimeters. The lesions remain intermittently associated with bowel loops including the medial aspect of the right colon. The lesion is also adjacent to the duodenum; however, there is no bowel obstruction or adjacent bowel wall thickening. It is noted to be adjacent to the liver with no lesions seen in the liver. The remainder of the exam was unremarkable. A. Coronal image B. Axial image.

with the most common presenting location being the head and neck, where lymphatic flow is rich.[1,4] Abdominal lymphatic malformations are much rarer, occurring at rates less than 5% that of all lymphatic malformations.[5]

Abdominal lymphatic malformations can involve the mesentery, omentum, gastrointestinal tract, and/or retroperitoneum. ALMs can present asymptomatically or with abdominal pain/distension, abdominal masses, acute obstruction, volvulus, and/or acute abdomen.[5] Often in children, the presenting symptoms are more acute with higher severity.[4] The mesentery is the most common site for ALMs and they are most commonly identified with CT/US, as in our case.[5] In general, ultrasound or MRI is better able to characterize abdominal lymphatic malformations.[4] With the presentation of a loculated cystic mass in the abdomen, the differential diagnosis includes uncomplicated ascites, abdominal tuberculosis, and malignant genitourinary masses.[5]

Previously, standard of care was surgical excision, especially in acute cases. **While resection is a surgical option, it carries significant potential morbidity due the extent and location of these conditions.** This has evolved due to the variable location of the malformation, diverse ages of patients, and availability of alternative treatment modalities. Currently, medical treatment and sclerotherapy are more attractive initial treatment steps, with sclerotherapy showing evidence of being able to avoid bowel resection, and systemic medical therapy with propranolol or sirolimus showing promise in cases where total excision of the malformation is not feasible.[4] While our patient's malformation seems to be diminishing in size, ALMs tend to recur and invade neighboring structures. Given the relative rarity of ALMs it has been difficult to define optimal clinical management. A modified classification system based on cyst size and predominance has shown promise in guiding therapy.[4] This case demonstrates the variability in presentation of ALMs and highlights the ease with which a large malformation might otherwise be undetected.

3. Conclusion

This unique case is a reminder of how large ALMs can present incidentally, disguised by other more common conditions, including a hydrocele or inguinal hernia. The case also illustrates the variability in natural history of abdominal lymphovascular malformations which can help inform clinicians when deciding on a treatment course. Depending on initial age of presentation and constellation of symptoms, more invasive treatment may be justified. Younger asymptomatic patients may benefit from watchful waiting. Careful follow-up is necessary given the potentially life-threatening complications; however, avoidance of ionizing radiation and invasive surgical interventions should be strongly considered.

Supplementary data related to this article can be found online at https://doi.org/10.1016/j.eucr.2021.101786

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