Mesenteric lymphatic malformation mimicking an adnexal source in a teenager

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Abstract

Mesenteric lymphatic malformations are rare benign tumors that are usually found in children and very rarely are they found in the abdomen. We present a rare case in which a mesenteric lymphatic malformation mimics an adnexal source. This is an important contribution to obstetrics and gynecology literature to show the importance of keeping this rare diagnosis on the differential, as well as the value of multi-disciplinary care.

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Introduction

Lymphatic malformations are rare congenital abnormalities that often present in the first few years of life, with 90% presenting in the first two years.[1] The majority of cases (95%) are located in the neck and axilla, with abdominal locations making up a mere few percent of the condition.[2] The exact prevalence of the condition is unclear, but it has been suggested in some literature that there may be as few as 820 cases of abdominal mesenteric lymphatic malformations since the 16th century.[1] Other literature indicates there have been less than 200 reports of mesenteric lymphatic malformations published in the English language literature.[3] The clinical picture varies widely, as some patients present with subacute diffuse abdominal discomfort, while others present with acute symptoms mimicking appendicitis.[1] Diagnosis cannot be made on the clinical examination exclusively, rather abdominal mesenteric lymphatic malformations are often found as an incidental finding on abdominal imaging. There have been only a few cases reported in which a mesenteric lymphatic malformation presents

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mimicking a pelvic mass or gynecologic emergency.[4-6] Here we present an atypical case of a mesenteric lymphatic malformation in a 15-year-old female, with uncharacteristic symptoms suggestive of an adnexal source.

Case Presentation

A nulliparous, menarchal, 44 kg, 15-year-old female presented to an outside hospital with lower abdominal pain for a week (left > right). The pain was constant and sharp, so she eventually went to her local Emergency Department (ED). There the patient denied symptoms of fever, vomiting, diarrhea, dysuria, hematuria, numbness, or tingling. She underwent a CT scan to rule out appendicitis and a loculated mass thought to be arising from her pelvis was found. The impression from the radiologist stated the mass appeared to be a cystic lesion in the abdomen and pelvis with a differential including an ovarian teratoma, venolymphatic malformation, or a cystadenoma. Due to concern that the mass could be malignant, the patient was transferred to the University of Iowa (U of I) Emergency Department for further evaluation.

Upon arrival, the patient's abdomen was soft, non-tender, and non-distended with normal bowel sounds. Her vital signs were within normal limits. Tumor markers for ovarian pathologies were obtained, including CA-125, AFP, CEA, and CA19-9; all returned within normal limits. She subsequently underwent a transabdominal pelvic ultrasound to further characterize the mass and to rule out ovarian torsion; this showed a large cystic structure in the pelvis surrounding the uterus and right ovary measuring 102 x 54 x 100mm. No blood flow was noted to the lesion. Given that the patient had no evidence of an acute abdomen, she was sent home with plans to return to the U of I Gynecology clinic the following week.

At the planned visit, the patient reported that the pain that resulted in imaging had resolved and it was determined that her prior pain was likely unrelated to the CT and ultrasound findings. However, given the uncertain etiology of the pelvic mass, the U of I Gynecology Oncology team was consulted. Upon review of her imaging, they consulted with Pediatric Radiology and together hypothesized that the abdomino-pelvic lesion was likely not ovarian in origin and more likely a malformation of the mesenteric lymphatics. A consultation with Pediatric Surgery at U of I was planned. Prior to that appointment, Pediatric Radiology requested a repeat transabdominal ultrasound, as well as an MRI, to further evaluate the mass prior to her visit Pediatric Surgery. These imaging studies produced results similar to the prior with multiple cystic areas throughout her lower abdomen, from the level of the umbilicus down to the pelvis, posterior to the bladder and the uterus. A few foci of calcification were seen. The SMA/SMV mesenteric vessels coursed through the center of the lesion. The mass was likely originating from a mesenteric lymphatic malformation (Image 1). Given the patient's stable clinical status and absence of symptoms, Pediatric Surgery recommended a non-operative approach with surveillance for any change in symptoms with scheduled surveillance imaging every 6 months.

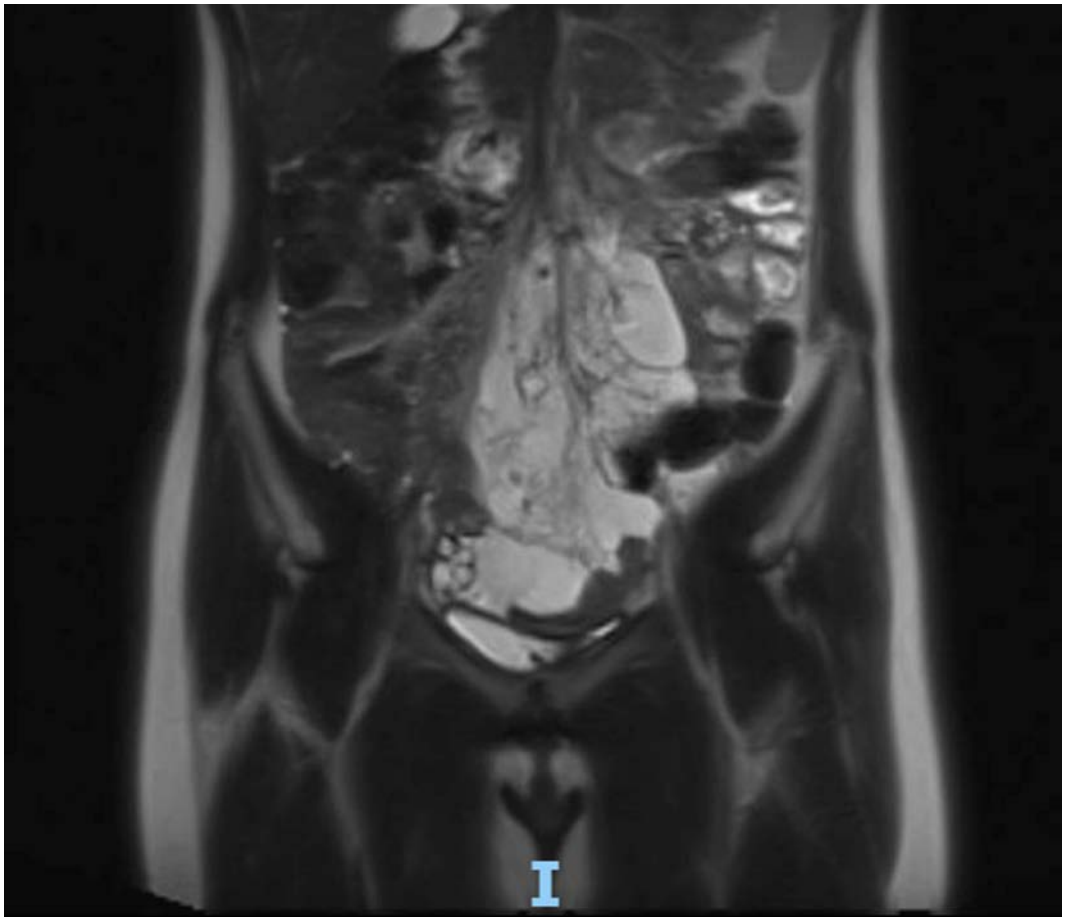

Image 1. Multicystic loculated lesion extending from retroperitoneum through mesenteric root to the pelvis.

Early during her surveillance, the patient was found to have occasional, mild, self-limiting abdominal cramping approximately every 2 weeks. Otherwise she continues to feel well, has normal bowel and bladder function, and normal menstrual cycles. Imaging continues to show a stable but stable, large, multicystic abdomino-pelvic lesion extending from the retroperitoneum through the mesenteric root to the pelvis, which Pediatric Radiology continues to believe is most consistent with a mesenteric lymphatic malformation.

Discussion and Conclusions

Mesenteric lymphatic malformations are rare benign tumors that are usually found in children.[7] The lymphatic system of the body is developed from endothelial channels in the neck, root of the mesentery, and the femoral and sciatic vein bifurcation.[8] Lymphatic malformations are likely caused by the presence of anomalous embryonic tissue that has inadequate drainage resulting in distension and discomfort.[4] They can be located in almost any part of the body, but very rarely do such

malformations arise in the mesentery.[3] Lymphatic malformations most often present clinically with gastrointestinal complaints such as nausea and vomiting. However, rarely they may be incidental findings due to imaging that was done for an alternative reason, as this case demonstrates. Diagnostically, it is very difficult to differentiate these lesions from other cystic intra-abdominal entities. On ultrasound, they often appear as multilocular cystic masses. Computed tomography can show homogenous fluid with low attenuation values.[8] These imaging findings overlap with many other possible intra-abdominal masses, so it can be difficult to make a definitive diagnosis based on imaging alone and not uncommonly, the diagnosis of a lymphatic malformation is not confirmed until histological specimens are examined after surgical intervention.[9] This case demonstrates the importance of keeping this diagnosis in the differential when an abnormal adnexal mass or pelvic abnormality is seen on CT or ultrasound. It also highlights the benefits of a multidisciplinary approach that may include Gynecology, General or Pediatric Surgery, Radiology, among other specialties.

Surgical management of lymphatic malformations is most often determined based on the cystic characteristics of the lesion. Macrocystic lesions are amenable to sclerotherapy, whereas microcystic lesions typically require resection. Surgical intervention is almost always curative if a complete resection is achieved.[1] In the patient presented, her lesion is essentially asymptomatic, and she is unlikely to achieve a complete resection considering the size and complexity of the mass. She additionally is not an optimal candidate for sclerotherapy due to the proximity of her complex lesion to surrounding great vessels. Our patient will continue to follow-up with her surgical team and if she develops symptoms, surgeons may consider laparoscopic unroofing of the large cysts. Risks of leaving this lymphatic malformation in place include the cyst becoming secondarily infected, a potential to invade other structures, and life threatening complications such as a volvulus or acute bowel obstruction.[1,3,9,10] However, in her current asymptomatic state, watchful waiting is deemed the most judicious approach.

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