



Massively enlarged wandering spleen with ruptured epidermoid cyst

© Muhammad Nur Syamim Che Johan¹, © Wan Zainira Wan Zain¹, © Andee Dzulkarnaen Zakaria¹, © Shafora Bibi², © Firdaus Hayati³

¹Universiti Sains Malaysia School of Medical Sciences, Department of Surgery, Kelantan, Malaysia

²Universiti Sains Malaysia School of Medical Sciences, Department of Radiology, Kelantan, Malaysia

³Universiti Malaysia Sabah Faculty of Medicine and Health Sciences, Department of Surgery, Sabah, Malaysia

Date submitted:

03.08.2023

Date accepted:

31.12.2023

Online publication date:

15.06.2024

Corresponding Author:

Wan Zainira Wan Zain, M.D.,
Universiti Sains Malaysia School of
Medical Sciences, Department of
Surgery, Kelantan, Malaysia
+097673000
zainira@usm.my

ORCID:

orcid.org/0000-0001-8019-6063

Keywords: Spleen, splenic cyst,
wandering spleen, splenectomy

ABSTRACT

Wandering spleen is extremely uncommon, with a reported incidence of <0.5%. A cyst within the spleen is also uncommon. The majority of primary splenic cysts fall into the epithelial cyst category. The affected patients present with atypical symptoms, like feeling fullness in the left upper abdomen and a palpable mass. The treatment of a large wandering splenic cyst is an open splenectomy. This case presents a unique combination of a wandering spleen with a huge ruptured cyst, featuring an unusual right lower abdominal and pelvic location, a twisted pedicle, and a remnant of normal splenic parenchyma, collectively constituting an exceedingly rare clinical scenario.

Introduction

The wandering spleen lacks normal attachments, connected by a vascular pedicle. Twisting can cause ischemia. Rarely, a splenic cyst occurs even rarer in a wandering spleen (1). They can be parasitic (hydatid), brought on by the parasite *Echinococcus granulosus*. The two main types of nonparasitic cysts are primary (epithelial), which are covered by an epithelial layer (epidermoid, dermoid, and mesothelial) or endothelial layer (hemangioma, lymphangioma), and secondary (pseudocysts, non-epithelial), which are typically the result of trauma (2). We here report a unique combination of a wandering spleen and a splenic cyst, delving into the potential causes, diagnostic challenges, and surgical approach.

Case Presentation

A 39-year-old nulliparous female with no known medical illness was admitted with a chief complaint of abdominal mass and generalized abdominal discomfort for 4 months. She reported increasing abdominal pain during the past 2 months. The mass had been growing gradually and was associated with vomiting episodes. She had no constitutional symptoms like weight loss, reduced appetite, or fever. Physical examination revealed a huge, hard tumor of approximately 20.0 x 15.0 cm (Figure 1A). Ultrasonography revealed an enlarged spleen of 16.0 cm in diameter. Moreover, a well-defined, large, anechoic lesion was detected in the top and mid poles of the spleen, with a remnant of normal splenic parenchyma at the lower pole.



There was very little echogenic moving debris, and no solid components or calcification (Figure 2A). Contrast-enhanced computed tomography (CT) scan of the abdomen showed that the spleen was situated abnormally in the right lower abdomen and pelvis. There was a large, clearly defined cystic lesion in the spleen measuring 12.8 x 17.5 x 14.7 cm (AP x W x CC) in size. The cystic mass within the wandering spleen showed the features of a ruptured cyst (Figures 2B, 2C).

An exploratory laparotomy and splenectomy were performed. Intraoperatively, a huge wandering spleen was observed, which occupied the abdomen with a twisted pedicle. The patient was stable postoperatively and was discharged home after 5 days of admission. The patient received oral penicillin and pneumococcal vaccinations as per the protocol post-splenectomy. Upon follow-up at 4 weeks, she was well without any surgical complications or recurrence. Histopathological examination of the cyst revealed a thin fibrous wall partly lined by squamous, transitional, and flat cuboidal epithelium, consistent with an epidermoid cyst.

Discussion

A normal spleen maintains its anatomical position by a few ligaments, including the gastrosplenic, lienorenal, and phrenocolic ligaments. A relatively uncommon disorder known as wandering spleen is characterized by the lack of all ligaments or the weakening of one or more of those ligaments (3). *Echinococcus granulosus* is a common type of parasitic splenic cyst.

The typical presentation of a wandering spleen is a mass with abdominal pain, but it may also present without any symptoms (4). The patient's primary concern was the presence of an abdominal mass. The patient described the mass as occupying a significant portion of her abdomen. In some patients, local inflammation of the left diaphragm may induce a chronic cough, and pressure in the cardiorespiratory system may cause pleuritic pain or dyspnea (5). Patients with acute torsion may exhibit acute severe abdominal pain, which often precedes fever and vomiting. In this instance, the patient could have experienced an acute-on-chronic presentation, given the gradual enlargement of the abdominal swelling over the preceding 4 months.

When examining chronic abdominal pain and the presence of an abdominal mass, it is important to include a splenic cyst as a possible consideration in the differential diagnosis. The unique features of splenic cysts on imaging help differentiate them from other conditions, such as ovarian cysts, gastrointestinal tumors, splenic tumors, or abdominal abscesses (6). For the acute abdomen, key biomarkers like whole blood count, serum lactate, lactate dehydrogenase, and blood urea nitrogen levels aid in early detection (7,8).

Ultrasonography is the preferred initial diagnostic method because it reveals the absence of the spleen in the left upper quadrant of the abdomen and indicates the presence of a solid mass in the abdomen or pelvis, consistent with our case. However, this method is operator-dependent, and bowel gas might obscure the findings (9). Typical CT findings for wandering spleen include the absence of the spleen anterior to the left

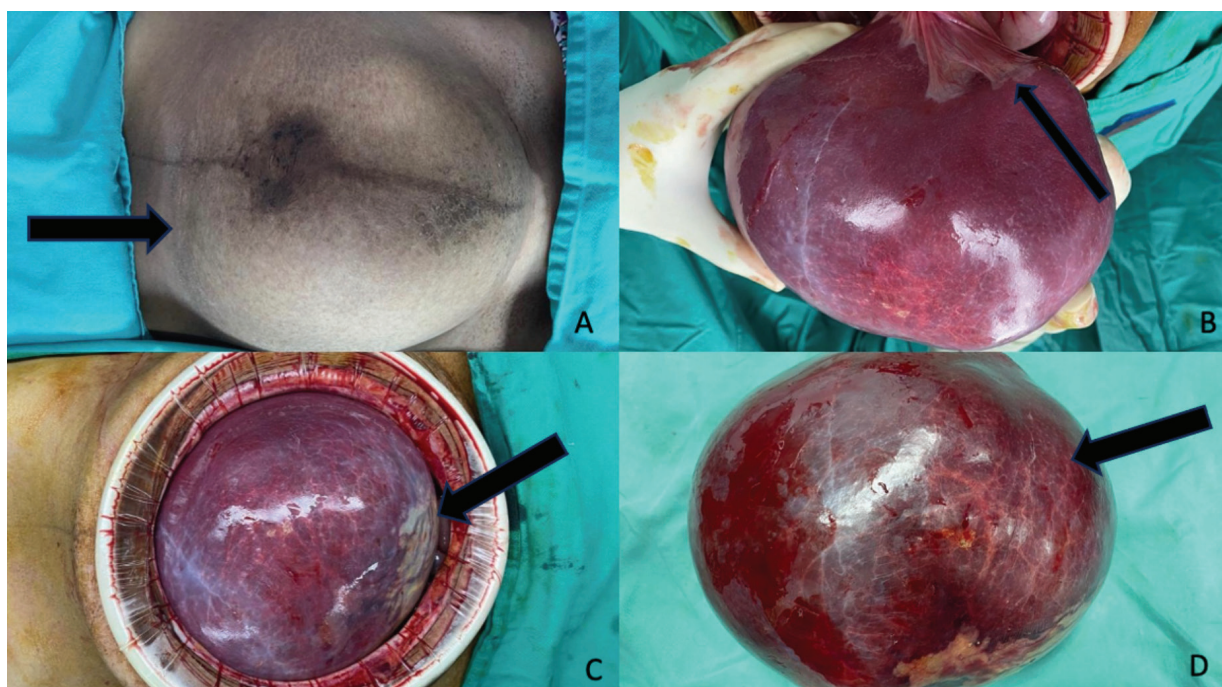


Figure 1. A) Abdominal examination revealed abdominal mass occupying the whole region of the abdomen and mimicking a termed size of the gravid uterus. B) A huge wandering splenic cyst with a twisted pedicle. However, the spleen was healthy and had no sign of ischemia/necrosis. C) A wandering spleen. D) Post-open splenectomy specimen showed wandering spleen with splenic cyst measuring 20 x 17 cm and weighed 2200 gm

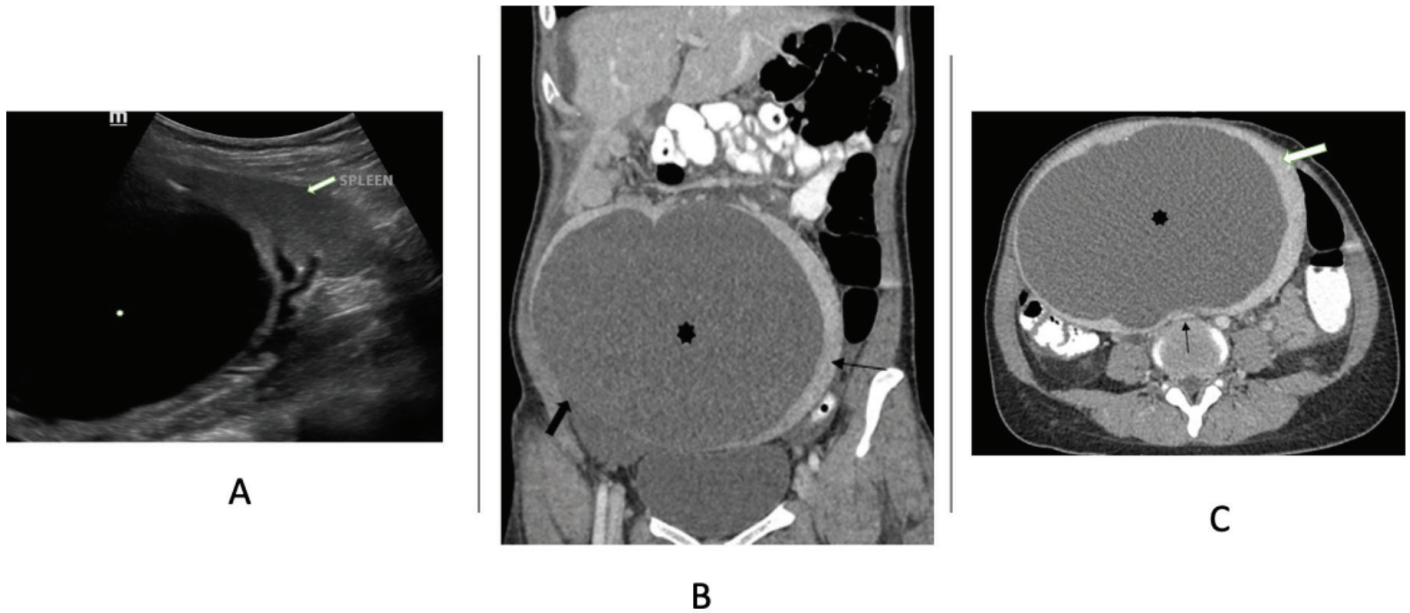


Figure 2. A) Ultrasound of the abdomen shows a large anechoic lesion (*) in keeping with the cyst occupying the spleen with remaining splenic parenchymal tissue (white arrow). B) Coronal view CT scan shows a large cyst (*) with the spleen (thin arrow). There is very thin remaining splenic parenchyma at the right lateral aspect with some areas showing focal discontinuity (big arrow) communicating with adjacent free fluid related to splenic rupture. C) Axial view CT scan shows spleen parenchymal tissue (big white arrow) at the lower abdomen with a large hypodense lesion (*) within in keeping with a cyst causing compression onto the left common iliac artery (thin black arrow). There is no solid component or septation within the cyst
CT: Computed tomography

kidney and posterior to the stomach with visualization of an abdominal or pelvic mass with homogenous or heterogenous splenic parenchymal enhancement (9). In this particular case, the spleen demonstrates regular enhancement without signs of splenic infarction. Nevertheless, a sizable cyst within the spleen exerts pressure on the splenic parenchyma and creates a mass effect on adjacent structures.

In the presented case, the cysts prompted clinical manifestations. Moreover, the CT image raises the suspicion of a ruptured cyst. As a result, surgical management was necessary. Treatment for a wandering spleen with a splenic cyst depends on cyst characteristics, patient health, and cyst size (10). Surgical options include open splenectomy or laparoscopic removal to preserve spleen function and manage associated risks. The laparoscopic approach is possible. Before the advent of laparoscopic surgery, primary splenic cysts are surgically managed with puncture, drainage, and marsupialization. However, splenic cysts can only be removed through laparoscopy if they are at the splenic poles or boundaries (11). Because of the risk of uncontrolled hemorrhages parenchymal cysts should not be drained. This is also applied for large splenic cysts where open splenectomy is the choice because of the possibility of significant complications (12). Considering our provisional diagnosis of a splenic cyst in a wandering spleen with a potential ruptured spleen, we opt for open splenectomy as the preferred surgical intervention (13).

Conclusion

In conclusion, this case emphasizes the need to consider rare conditions like wandering spleen with a cyst in patients with abdominal masses and pain. Accurate diagnosis, aided by advanced imaging, enables effective surgical intervention and enhances patient care and medical insights.

Ethics

Informed Consent: Informed consent was provided by the participant.

Authorship Contributions

Surgical and Medical Practices: M.N.S.C.J., W.Z.W.Z., S.B., Concept: W.Z.W.Z., Design: F.H., Analysis or Interpretation: A.D.Z., Literature Search: S.B., Writing: M.N.S.C.J., F.H.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Falchetti D, Torri F, Dughi S, et al. Splenic cyst in a wandering spleen: laparoscopic treatment with preservation of splenic function. *J Pediatr Surg.* 2007;42:1457-1459.
2. Reddi VR, Reddy MK, Srinivas B, Sekhar CC, Ramesh O. Mesothelial splenic cyst--a case report. *Ann Acad Med Singap.* 1998;27:880-882.

3. Varga I, Babala J, Kachlik D. Anatomic variations of the spleen: current state of terminology, classification, and embryological background. *Surg Radiol Anat.* 2018;40:21-29.
4. Göksu M, Baykan AH. Torsion of Wandering Spleen: A Case Report. *J Emerg Med.* 2020;58:189-192.
5. Labruzzo C, Haritopoulos KN, El Tayar AR, Hakim NS. Posttraumatic cyst of the spleen: a case report and review of the literature. *Int Surg.* 2002;87:152-156.
6. Vancauwenberghe T, Snoeckx A, Vanbeckevoort D, Dymarkowski S, Vanhoenacker FM. Imaging of the spleen: what the clinician needs to know. *Singapore Med J.* 2015;56:133-144.
7. Verma I, Kaur S, Goyal S, Goyal S, Multani JS, Narang AP. Diagnostic value of lactate levels in acute abdomen disorders. *Indian J Clin Biochem.* 2014;29:382-385.
8. Uçaner B, Buldanlı MZ, Çiftçi MS, Çimen Ş, Hançerlioğulları O. Are Mortality and Morbidity Predictable in Acute Mesenteric Ischemia? The Importance of Serum Lactate, Lactate Dehydrogenase, and Blood Urea Nitrogen Levels. *Indian J Surg.* 2023;85:1191-1197.
9. Faridi MS, Kumar A, Inam L, Shahid R. Wandering Spleen- A diagnostic Challenge: Case Report and Review of Literature. *Malays J Med Sci.* 2014;21:57-60.
10. Ikeda T, Hosokawa T, Goto S, Hashimoto M, Nagasaki E, Masuko T. Successful laparoscopic-assisted partial splenectomy and splenopexy with umbilical approach to wandering spleen with an enlarged cyst in a pediatric patient. *J Surg Case Rep.* 2022;2022:rjac483.
11. Yoh T, Wada S, Kobayashi A, et al. Laparoscopic splenectomy for a large multilocular splenic cyst with elevated CA19-9: Report of a case. *Int J Surg Case Rep.* 2013;4:319-321.
12. Hui Lian H, Hayati F, Ali AA, et al. Wandering spleen: a unique cause of acute abdomen. *Folia Morphol (Warsz).* 2018;77:400-402.
13. Najid F, Sanjeev S, Zuki A, et al. Wandering Spleen as a Cause of Acute Abdomen: A Surgical Conundrum from Acute Appendicitis to Splenic Torsion and Ischemic Small Bowel Volvulus. *Malays J Med Sci.* 2020;16:336-338.