Case Report

Wandering Spleen containing Mesothelial Cyst: A case report and review of literature

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ABSTRACT

Wandering spleen, defined as a spleen without peritoneal attachment, is very rare and the presence of a cyst in it makes it even rarer. We report a case of a wandering spleen containing a mesothelial cyst. The case presented with recurrent lower abdominal pain. A CT scan was diagnostic, and laparoscopic assisted splenectomy was performed.

KEY WORDS: cyst, splenectomy, wandering spleen

INTRODUCTION

The spleen develops from the mesoderm in the dorsal mesogastrium. It lies in the left hypochondrium behind the stomach, and is approximately 12 cm long and 7 cm wide[1]. The spleen is fixed by five ligaments or peritoneal reflections, which are embryological condensations[2]. Wandering spleen, defined as a spleen without peritoneal attachment, is a rare entity with an incidence of less than 0.5%[3]. Splenic cysts are classified as parasitic and non-parasitic cysts, the non-parasitic are further categorized as primary (epithelial-true) and secondary (pseudo-false) cysts based on the lining of the cyst[4]. We report a case of a wandering spleen containing a mesothelial cyst, discussing the possible ways of management.

CASE REPORT

A 17-year-old female was referred to our surgical outpatient clinic with a few days history of occasional dull aching left and lower abdominal pain with no associated symptoms. On examination, a painless and partially mobile pelvi-abdominal mass was felt, occupying mainly the suprapubic region. An ultrasonography of the abdomen was performed and showed a pelvi-abdominal mass containing a unilocular cystic mass measuring 10 x 9 x 9.8 cm with turbid internal echoes. The impression was most likely pelvic spleen with a cyst within it (Fig 1). The patient was admitted to the hospital for further workup. Her laboratory tests were within normal limits. A CT scan of the abdomen with contrast enhancement showed a wandering spleen measuring 19.3 x 14.6 cm with a large splenic cyst in the centre measuring 9.7 cm with a very thin wall (Fig 2). The case was discussed with the patient and her family and they agreed for splenectomy. After receiving anti-pneumococcal, Hemophilus influenza and meningococcal vaccines, the patient underwent laparoscopic assisted splenectomy via 3 ports. A 10 mm supra-umbilical port was used beside a 10 mm port in the right mid-clavicular line just above the level of the umbilicus and a 5 mm port at the McBurney's point. The spleen was found in the pelvis with fine ligament attachments. Splenic pedicle was dissected and separated by staplers, the freely mobile spleen was fitted into an endo-bag and extracted through the supra-umbilical port site after enlargement of the incision (Fig 3). The patient tolerated the procedure well and was discharged two days later. The histopathological examination revealed a mesothelial type of splenic cyst (Fig 4). After more than three months follow-up, the patient is free of abdominal pain.

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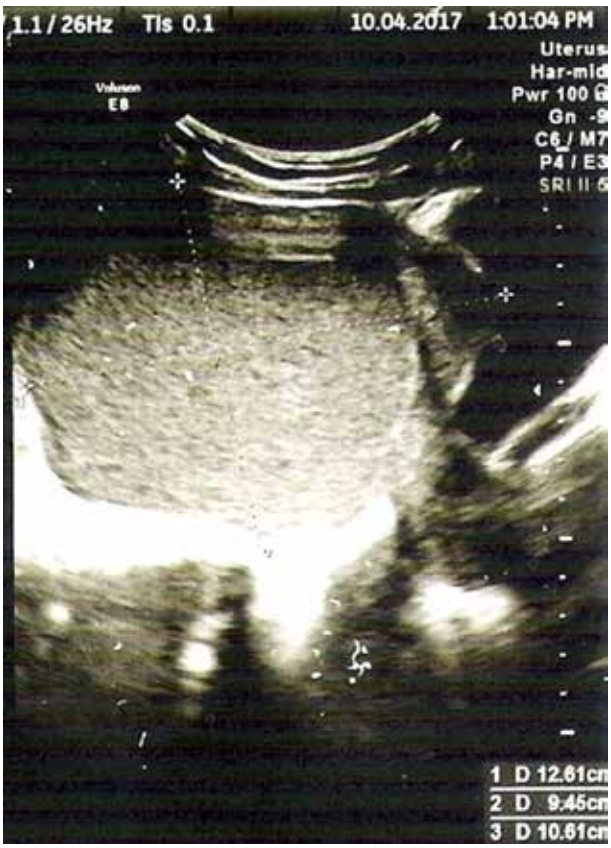

Fig 1: Ultrasonography display of the abdomen and pelvis showing the wandering pelvic spleen and a large splenic cyst.

DISCUSSION

The first description of a wandering spleen is attributed to Von Horne in 1667, as an autopsy finding on an adult[5]. Wandering or ectopic spleen has two possible aetiologies: congenital and acquired. The congenital form occurs due to failure of the dorsal mesogastrium to develop when the lesser sac is formed. The acquired form occurs mostly in multiparrous female, as the ligament, which holding the spleen in position become lax[6]. The male to female ratio is about 1:7 in adults and most females are in the child-bearing age group of 20 to 40 years[3]. Wandering (ectopic or floating) spleen as an entity is very rare and the presence of a cyst in it makes it even rarer[7]. The splenic cysts are classified as parasitic and non-parasitic cyst. Parasitic cysts are generally seen in endemic areas and are usually caused by Echinococcus granulosus infestations. The true or primary non-parasitic cysts maybe congenital or neoplastic in origin and are lined by mesothelial, squamous or transitional epithelium. Secondary or pseudo non-parasitic cysts are usually post-traumatic, due to failure of organization of sub-capsular or parenchymal hematomas[8]. Non-parasitic cysts of the spleen are rare and are incidentally discovered, derived from the mesothelial cell lining of the splenic capsule[9]. Embryonic inclusion of epithelial cells from adjacent structure, epithelial cell metaplasia from adjacent structures or vascular endothelium from peritoneal inclusions are some of the theories to explain the genesis of these congenital cysts[9]. Also, capsular surface mesothelial invagination results in subsequent cyst formation[4]. Primary true cysts of the spleen account for about 10% of all non-parasitic cysts of the spleen[10]. Mesothelial cysts have a characteristic gross and microscopic appearance[9]. Grossly, they are usually unilocular and vary in size, with whitish or greyish-white cut surface[11]. Microscopically, the lining cells stain positive for calretinin, a mesothelial marker,

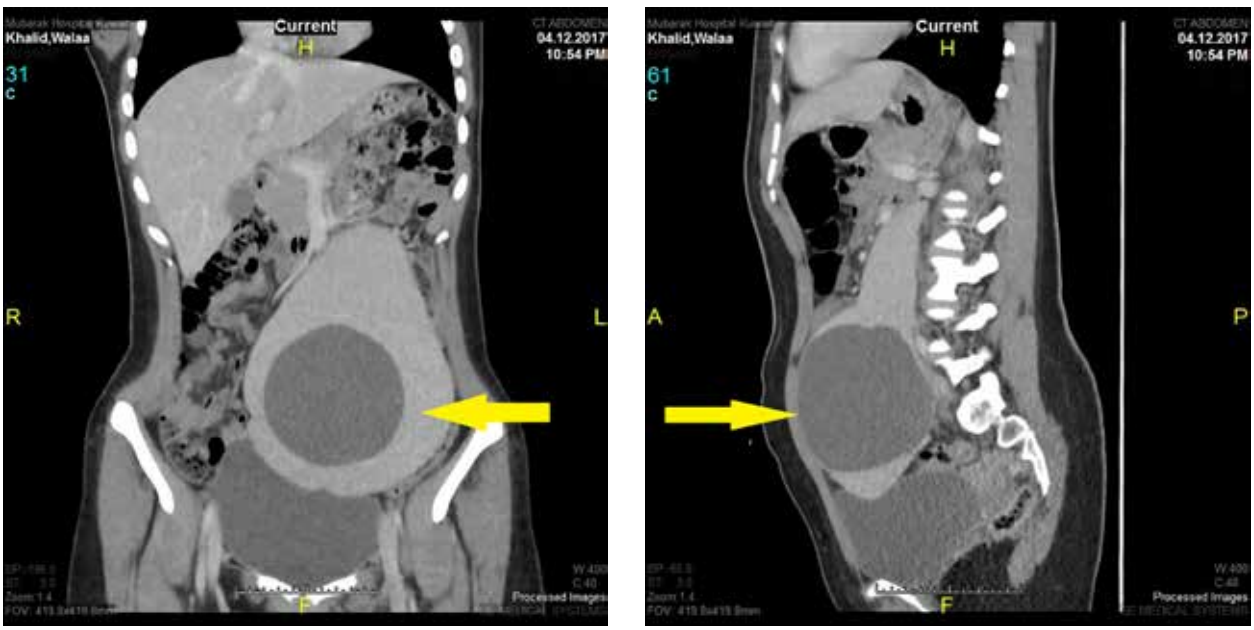

Fig 2 (A&B): Computed tomography scan abdomen (A) coronal (B) sagittal view, showing a large cyst in a wandering pelvic spleen.

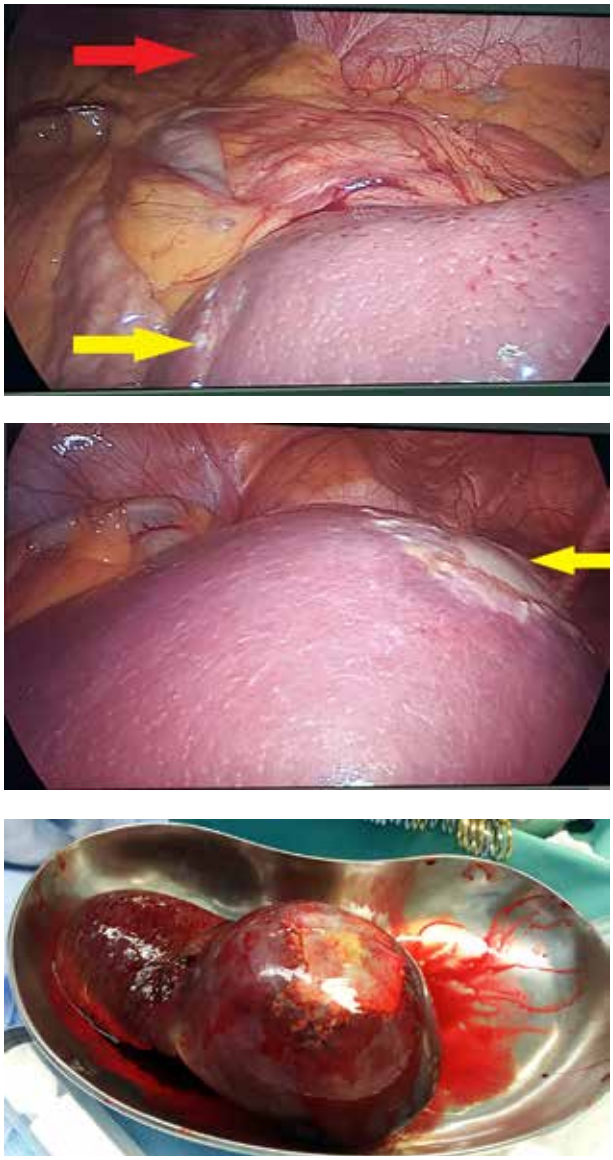

Fig 3 (A,B&C): Intra-operative picture (A) laparoscopic view with the yellow arrow pointing to the pelvic spleen and the red arrow pointing to normal anatomical site (B) the pelvic spleen with the cyst (C) the extracted spleen.

and secrete CA 19-9 which can be used as a screening tool in the management of these lesions[12]. Studies have indicated a rise in CA 19-9 in association with primary mesothelial cyst and the reduction of its level after cyst removal, offering a screening test to indicate recurrence in case of spleen preserving surgeries[11]. Clinical presentation of these cases (wandering spleen with or without a cyst) usually present with an asymptomatic abdominal mass or a mass associated with recurrent pain, also may present as an acute abdomen if it becomes complicated by torsion[13]. Wandering spleen also can predispose to other life-threatening complications such as splenic infarction, portal hypertension or gastrointestinal bleeding[14]. Imaging studies such as a colour ultrasonography, visceral arteriography, splenic radionucleotide scan and contrast enhanced spiral CT are very useful in reaching a definitive diagnosis of a wandering spleen[15]. Regarding mesothelial cyst, the preoperative diagnosis is rare, but they can be suspected in case of unilocular cysts with no previous history of trauma, infection, or exposure to hydatid infection. Imaging may be useful in the investigation, such as ultrasound of the abdomen, which shows whether the cysts are

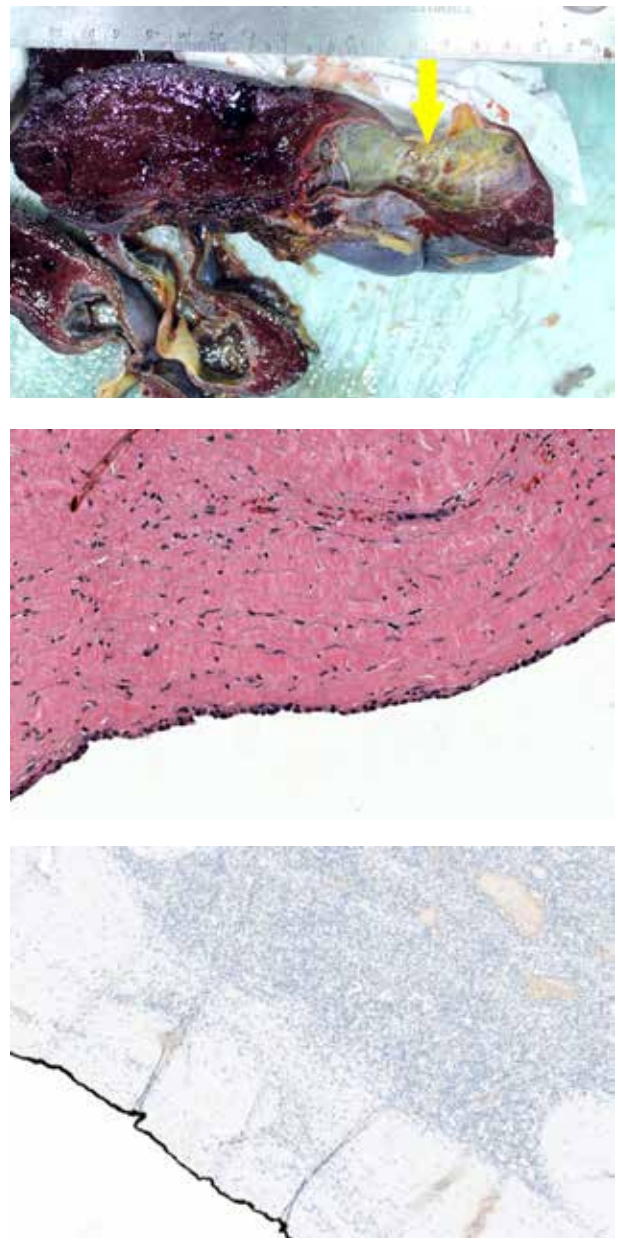

Fig 4 (A,B&C): Histopathological examination of the splenic cyst (A) cut section showing a single large fibrous trabeculated cyst (B) microscopy showing the cyst wall lined by cuboidal mesothelium (H & E x100) (C) cyst epithelial lining showing positivity for calretinin (immunohistochemistry x200).

anechoic or hypoechoic, in addition to showing wall thickness. Computed tomography scans (CTs) and magnetic resonance imaging, can provide additional information regarding the morphology of the cyst, the composition of the cyst fluid, the precise location of the cyst in the spleen, and its anatomical relationship with the surrounding abdominal organs. The diagnosis is confirmed histopathologically[16]. Splenectomy has traditionally been used for wandering spleen, splenopexy is increasingly used in the pediatric population to anchor the spleen and preserve splenic function. Concerns over overwhelming post-splenectomy sepsis make splenopexy the first line of treatment if there was no evidence of infection or any other complicating pathology[17]. However, a multicentric study reported complications after salvage with splenopexy in 60% of cases resulting in post splenopexy splenic ischemia[18]. Regarding splenic cyst, conservative treatments with regular scans may be an option for cysts that are up to 5 cm in diameter, completely asymptomatic, and which exhibit the most typical characteristic of non-parasitic splenic cysts. In cases with surgical indication and due to the growing population of spleen preservation to preserve immune function, many alternative treatment options have been suggested[10,19]. Also, surgery is the gold standard treatment for symptomatic and large (>5 cm) splenic cysts and the surgical procedure is tailored depending on the patient's age and the size, nature and location of the cyst. The various surgical procedures described are open complete splenectomy, partial splenectomy, cystectomy, marsupialization and cyst decapsulation[20]. Laparoscopic management of splenic cysts offers the advantage of minimally invasive surgery, faster recovery, shorter hospital stay, and reduced morbidity. Anterior surface splenic cysts are more amenable to laparoscopic fenestration than the posterior surface cysts as greater splenic mobility is needed in the latter. Open partial or complete splenectomy is advocated for centrally located splenic cysts[21]. Anti-pneumoccocal, hemophilus influenza, and meningococcal vaccines are indicated before elective splenectomy and shortly after non-elective splenectomy[22]. In our case, the wandering spleen was diagnosed by ultrasonography and confirmed by a CT scan. The aetiology of the spleen was most likely congenital, but it could be acquired due to the lax and fine ligament attachments found intra-operatively as a result of the large cyst. The splenic cyst was diagnosed as a primary cyst due to the age of the patient, the negative history of any previous infectious or exposure to hydatid infection, and the finding of unilocular cysts in both the ultrasonography and the CT scan. After receiving the vaccinations, a laparoscopic assisted total splenectomy was performed. We preferred total splenectomy as it is indicated in case of wandering spleen and due to the presence of a large central cyst.

CONCLUSION

Wandering spleen as an entity is very rare and the presence of a cyst within it makes it even rarer. A laparoscopic approach for splenectomy in such cases is an option, but laparoscopic assisted approach is much safer, especially in case of a large splenic cyst. We would like to draw attention to this rare entity in the differential diagnosis of an abdominal mass with minimal symptoms.

REFERENCES

1. Singh I. Essentials of anatomy. New Delhi: Jaypee Brothers Medical Publishers Ltd; 2002. p 283.
2. Kanthan R, Radhi JM. The "True" splenic wander. Can J Gastroenterol 1999; 13(2):169-171.
3. Kapan M, Gümüs M, Onder A, Gümüs H, Aldemir M. A wandering spleen presenting as an acute abdomen: Case report. J Emerg Med 2012; 43(5):e303-305.
4. Ough YD, Nash HR, Wood DA. Mesothelial cysts of the spleen with squamous metaplasia. Am J Clin Pathol 1981; 76(5):666-669.
5. Correnson H. Quelques Recherches Sur les Displacements de Larate. Paris: A Parent, 1876.
6. Alawi MH, Khalifa A, Bana SH. Wandering spleen: a challenging diagnosis. Pak J Med Sci 2005; 21(4):482-484.
7. Labruzzo C, Haritopoulos KN, ElTayar AR, Hakim NS. Post traumatic cyst of the spleen: a case report and review of the literature. Int Surg 2002; 87(3):152-156.
8. Mirilas P, Mentessidou A, Skandalakis JE. Splenic cysts: are there so many types? J Am Coll Surg 2007; 204(3):459-465.
9. Kuwabara S, Hohjoh H, Nakano M, *et al*. Mesothelial splenic cyst. Intern Med 1993; 32:672-674.
10. Morgenstern L. Nonparasitic splenic cysts: pathogenesis, classification, and treatment. J Am Coll Surg 2002; 194(3):306-314.
11. Higaki K, Jimi A, Watanabe J, Kusaba A, Kojiro M. Epidermoid cyst of the spleen with CA 19-9 or Carcinoembryonic antigen production: report of three cases. Am J Surg Pathol 1998; 22(6):704-708.
12. Trompetas V, Panagopoulos E, Priovolou-Papaevangelou M, Ramantanis G. Giant benign true cyst of the spleen with high serum level of CA 19-9. Eur J Gastroenternal Hepatol 2002; 14(1):85-88.
13. Steinberg R, Krmazyn B, Bdugy E, Gelber E, Freud E, Horev G, *et al*. Clinical presentation of wandering spleen. J Pediatr Surg 2002; 37(10):E30.
14. Chan KC, Chang YH. Acute abdomen due to torsion of a pelvic wandering spleen. J Formos Med Assoc 2002; 101(8):577-580.
15. Wen YL, Kudo M, Maekawa K, Momuwa H, Haji S, Ohyanagi H. Sonographic findings of wandering spleen. J Gastro Enteral 2001; 36:693-694.

16. Zvizolic' Z, Karavolic' K. Spleen-preserving surgery in treatment of large mesothelial splenic cyst in children - a case report and review of the literature. Bosn J Basic Med Sa 2013; 13:126-128.
17. Mc Heik JN, Richer JP, Levard G. Torsion of the spleen in children. Arch Pediatr 2005; 12:1496-1499.
18. Fiquet - Francois C, Belouadah M, Ludot H, *et al*. Wandering spleen in children: multicenter retrospective study. J Pediatr Surg 2010; 45:1519-1524.
19. Vijayaraghavan R, Chandrashekar R, Aithal S, Rashmi MV, Belagavi CS. Mesothelial cyst of the spleen in an adult: a case report. BMJ case Rep 2010; 2010:bcr0320102810.
20. Reddi VR, Reddy MK, Srinivas B, Sekhar CC, Ramesh O. Mesothelial splenic cyst - a case report. Ann Acad Med Singapore 1998; 27(6):880-882.
21. Geraghty M, Khan IZ, Canlon KC. Large primary splenic cyst: A laparoscopic technique. J Minim Access Surg 2009; 5(1):14-16.
22. Benevento A, Boni L, Dionigi G, Ferrari A, Dionigi R. Emergency laparoscopic splenectomy for "wandering" (pelvic) spleen: case report and review of literature on laparoscopic approach to splenic disease. Surg Endosc 2002; 16(9):1364-1365.