Case Report

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

Khalid H Al-Hammad, Adel Al-Fudari, Mohammed Ibrahim
Kuwait Board of Surgery
Department of Surgery, Mubarak Al-Kabir Hospital, Kuwait

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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.

Address correspondence to:
Dr. Khalid Hamad Al-Hammad, Department of Surgery, Mubarak Al-Kabir Hospital, Al-Jubriya, P.O.Box 43787, Code: 32052 Kuwait. Tel: (+965)99108191; E-mail: duke_alhammad@hotmail.com
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 multiparous 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 subcapsular 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\[9\]. 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,
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[13]. 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 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.

**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. Splenectomy has traditionally been used for wandering spleen, splenectomy is increasingly used in the pediatric population to anchor the spleen and preserve splenic function. Concerns over overwhelming post-splenectomy sepsis make splenectomy the first line of treatment if there was no evidence of infection or any other complicating pathology. However, a multicentric study reported complications after salvage with splenectomy in 60% of cases resulting in post splenectomy splenic ischemia. 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. 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. Laparoscopic management of splenic cysts offers the advantage of minimal 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. Anti-pneumococcal, hemophilus influenza, and meningococcal vaccines are indicated before 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


