A congenital wandering spleen with a large epithelial cyst: A case report

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Abstract
We present a case of a 21-year-old female patient who had a wandering spleen with a large cyst. The celiotomy was performed under the impression of torsion of the wandering spleen and leakage from the splenic cyst. The total splenectomy was performed instead of cystectomy and splenopexy.

KEYWORDS
splenectomy, splenic epithelial cyst, splenic true cyst, wandering spleen

1 | INTRODUCTION

The wandering spleens and splenic cysts are rare diseases, respectively. Wandering spleens result from the elongation or underdevelopment of suspensory ligaments in the spleen. It usually occurs in children and among adults, it is commonly found in females of active reproductive age. Wandering spleens may cause intermittent abdominal discomfort due to torsion and spontaneous detorsion of the spleen but usually remains an asymptomatic mass in the abdomen. True cysts containing cellular epithelial lining consist of about 20% of all splenic cysts. The clinical presentation of splenic cysts can vary, ranging from being incidentally discovered on routine imaging to nausea, vomiting, vague abdominal pain, or rarely, painful splenomegaly, particularly when the cyst is large.

So far, a small number of cases of wandering spleen accompanying splenic cyst have been reported. And only 5 cases among them had the wandering spleen with splenic true cyst. We report a case of a 21-year-old woman having a congenital wandering spleen with a congenital epithelial cyst.

2 | CASE REPORT

A 21-year-old Cambodian woman presented with intermittent abdominal pain and a palpable abdominal mass in the left lumbar area for about 1 year. The abdominal pain and the size of the mass had progressively increased a few months before visiting us. Sometimes, the patient experienced nausea, vomiting, and bloating. On physical examination, she had a palpable mass in the left paraumbilical region, and it was about 11 cm in size and tender, mobile, and had a smooth surface and well-defined margins. There was also diffuse and mild tenderness with a rebound on the whole abdomen.

The laboratory parameters showed mildly elevated WBC (11,700 per μl) and neutrophil count (78.1%), and hemoglobin was 11.8 g/dL with low mean corpuscular volume (61.5 fl) and low mean corpuscular hemoglobin (19 pg). Other blood and urine laboratory results were within normal limits.

On images of computer tomography (CT), the spleen with cyst was positioned from the left hypochondrium to the
left lower quadrant of the abdomen, the size of the spleen and cyst was approximately 10 cm, respectively (Figure 1). The splenic pedicle was rotated and looked like an “archery target” on the transverse view (Figure 2). The intra-abdominal fluid collection was suspected to be the result of fluid leakage from the splenic cyst. Therefore, we suspected that the leakage from the cyst in the wandering spleen had resulted in the symptoms and signs of mild peritonitis.

A celiotomy was performed using a mid-midline incision. The splenic pedicle and mesentery were long and rotated in a counterclockwise direction; however, the rotation was slack and there was no infarction of the spleen (Figure 3). The size of the splenic cyst was about 10 cm, and the cystic wall was wet but seemed like intact without rupture or a hole. There was approximately 200 cc of fluid collection in the abdominal cavity, which was clear and thin. And we removed the spleen with the cyst without the biopsy for the frozen section of the cyst wall because we do not have the facility for the procedure.

On the pathology report, the size of the spleen was 17×11×7 cm, and the size of the cyst was 11×10×7 cm (Figure 4). Grossly the cyst contained yellowish-brown fluid and had a glistening inner surface with white trabeculations and there was no evidence of rupture or hole in the cyst wall. Microscopic findings revealed that the cyst wall was lined by cuboidal or mesothelial-like epithelium with focal calcification and adnexa of skin was not observed (Figure 5). There were no atypical cells or evidence of malignancy in the specimen of the splenic cyst. Inflammatory cells infiltrated the pedicle of the spleen, but the necrosis of the spleen was not observed.

3 | DISCUSSION AND CONCLUSION

A wandering spleen is a rare clinical entity with an incidence of less than 0.2%. Congenital wandering is caused by failure of complete fusion of the mesogastrium and posterior abdominal wall during the second month of pregnancy, which may be the absence or maldevelopment of one or more suspensory ligaments (gastroplenic, splenorenal, splenocolic, splenophrenic, and pancreaticosplenic ligaments). The abnormal ligamentous laxity fails to fixate the spleen in its normal location in the left upper quadrant.

Cystic lesions of the spleen can be parasitic or non-parasitic. Nonparasitic cysts are of two types: primary cysts and secondary pseudocysts. Primary cysts of the spleen are very rare and are also called true, congenital, epidermoid, or epithelial cysts. The incidence rate of splenic cysts is very low (0.07%) in patients who undergo autopsies. The incidence rate of splenic cysts has increased up to 1% because of the broad use of ultrasonography, CT, and magnetic resonance imaging. Davidson and colleagues documented the first splenic cyst in an ectopic spleen in an article in 1980. True cysts containing a cellular epithelial lining comprise approximately 20% of all splenic cysts. Congenital cysts are assumed to be derived from peritoneal invasion, with the mesothelial lining accompanying the capsule during development. The pluripotent nature of the cellular lining tends to lead to metaplasia, fluid secretion, and cyst development.

Wandering spleens accompanying cysts are rarer than the respective disease of wandering spleen and splenic cyst. So far, 7 cases of wandering spleen with splenic cyst have been reported in a review of the literature (Table 1). But only 5 cases among them had the wandering spleen with splenic true cyst. Manoharan et al. reported a wandering spleen with a cyst, but the cyst was a pseudocyst. And Nawaz et al. also reported a wandering spleen with a cyst in a neonate, and they did partial splenectomy and splenopexy. But the splenic cystic degeneration was caused by infarction of the lower pole of the spleen due to intrauterine torsion of splenic vessels.

The mechanism underlying the relationship between the wandering spleen and the splenic cyst is not well understood. There may be a concurring invasion of the
epithelial lining in utero during the perinatal period, which progresses to epithelial cysts and may coexist with abnormal development of ligaments leading to the congenital wandering spleen.\textsuperscript{6,7,9,10,12}

History taking and physical examination are the primary tools for making an initial impression. Some patients may be asymptomatic or complain of mild intermittent pain because splenic congestion varies with intermittent torsion and spontaneous detorsion.\textsuperscript{7,9,12} Abdominal ultrasonography or CT is the primary imaging modality.\textsuperscript{11} CT contrast imaging is very helpful to identify the morphology of cysts and abnormal position of the spleen, with or without torsion in the abdomen or pelvis. The splenic pedicles looking like “archery target” are specific for splenic torsion (Figure 2).

A wandering spleen can become potentially life-threatening if there is a delay in the clinical or radiological diagnosis, leading to splenic torsion, infarction, and necrosis.\textsuperscript{20} The detorsion of the splenic pedicle and splenopexy using the laparoscopic approach is a possible surgical choice of treatment when there is no evidence of splenic infarction.\textsuperscript{21} For the management of splenic cysts in the wandering spleen, some authors reported that they did laparoscopic cystectomy and splenopexy (Table 1).\textsuperscript{4,5} Falchetti et al. reported a non-parasitic cyst in a wandering spleen in a 14-year-old girl and performed laparoscopic unroofing of the cyst and splenopexy in a vycril mesh (Table 1).\textsuperscript{6} On the contrary, others did total splenectomy by open approach or laparoscopic assistance (Table 1).\textsuperscript{7–9} But the various types of conservative surgery are of less value in intrasplenic cysts almost surrounded by splenic parenchyma, multiple cysts, and large cysts in the hilum of the spleen, with dense vascular adhesions to surrounding structures.\textsuperscript{22} In such circumstances, splenectomy either by open approach or by laparoscopic approach is the treatment of choice. In our case, we decided to do a total splenectomy because the total excision of the cyst or partial splenectomy with splenopexy was not available in our circumstances, and we were not confident in bleeding control without energy or a laser device. After splenectomy, our patient received only the pneumococcal vaccine (Prevenar 13) because the vaccines for the Meningococcus and Haemophilus influenzae are not available in our situation.

\textbf{FIGURE 2} The splenic pedicle was rotated and looked like an “archery target” on the transverse view of CT scan

\textbf{FIGURE 3} The wandering spleen and splenic cyst after detorsion of the pedicle

\textbf{FIGURE 4} The gross findings of the wandering spleen and splenic cyst
AUTHOR CONTRIBUTIONS
YL is the first author, and YDL is the corresponding author. VC as a pathologist described the pathologic findings of the specimen. HL and CHL contributed to the collection of the figures. All authors reviewed the manuscript.

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CONFLICT OF INTEREST
The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT
Data available on request due to privacy/ethical restrictions.

ETHICAL APPROVAL
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CONSENT
Written informed consent was obtained from the patient for publication.

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FIGURE 5 The microscopic findings of the splenic cyst wall lined by cuboidal epithelium or mesothelial-like cells (H&E 100 magnification)

TABLE 1 Reported Cases of the Congenital Wandering Spleen accompanying Splenic Cyst

| Category                      | Author       | Operation Method                      |
|-------------------------------|--------------|---------------------------------------|
| Wandering spleen with a true cyst | Prashant U 2007 | Laparoscopic cystectomy and splenopexy^4 |
|                               | Baglaj M 1998 | Laparoscopic cystectomy and splenopexy^5 |
|                               | Falchetti D 2007 | Laparoscopic unroofing of the cyst and splenopexy^6 |
|                               | Awe JAA 2005 | Splenectomy^7                          |
|                               | Kum CK 1993  | Splenectomy with laparoscopic assistance^8 |
| Wandering spleen with a pseudocyst | Manoharan D 2019 | Splenectomy^9 |
|                               | Nawaz A 2000 | Partial splenectomy and splenopexy^10 |

(A) (B)
presenting as an abdominal cystic swelling. *J Pediatr Surg.* 2000;35(10):1508-1510.

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