

Epidermal splenic cyst

G. H. M. Pinsara, N. Liyanage, P. G. K. Anuradha, P.M. Lambiyas, H. Gamage
Gastrointestinal Surgical Unit, Teaching Hospital Karapitiya, Galle.

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Abstract

We report a case of a large epithelial cyst of the spleen of an 18 year old man who was treated with splenectomy. The management of splenic cysts has changed from total splenectomy to splenic preservation surgery. But complete splenectomy is reserved for cases in which cyst excision cannot be done.

Introduction

Splenic cysts are rare lesions with 800 cases reported in the world literature [1]. Splenic cysts are classified into primary (true) or secondary (false) cysts on the basis of presence or absence of cellular lining of the cystic wall [2].

The true incidence of splenic cysts is unknown. The prevalence of splenic cysts has increased recently secondary to increased detection with the computerized tomography and the non-operative management of certain types of splenic injury [3]. Here we present a case of non-parasitic large epithelial cyst.

Case report

An 18 year old man presented with left sided abdominal distension for 4 years. There was no history of abdominal pain, history of trauma, significant medical illness or surgical intervention in the past. Abdominal examination revealed a large mass in the left hypochondrium extending to the umbilical region and epigastrium.

The patient had a contrast enhanced CT scan of the abdomen which showed a large cystic lesion measuring 20×20×21cm in relation to the pancreas. Based on imaging, the following diagnoses were considered;

1. A cyst arising from the pancreas

2. A cyst arising from the spleen

In order to determine whether it was a pancreatic cyst or a splenic cyst an ultrasound guided aspiration of the cyst was arranged and the aspirate was sent for amylase, carcino-embryonic antigen (CEA), CA 19-9 and cytology. Cytology revealed a straw coloured proteinaceous fluid with no evidence of micro-organism growth or malignant cells. CEA, CA 19-9 and amylase levels were normal. Other haematological investigations were unremarkable. This makes the diagnosis more in favour of a cyst arising from the spleen. Exploratory laparotomy was planned.

As there was a possibility of a splenectomy he was referred to the haematology unit and he was given polyvalent pneumococcal and meningococcal vaccines.

At laparotomy a large splenic cyst was found in relation to the hilum of the spleen. The spleen was elongated, flattened and was sandwiched between the anterolateral abdominal wall and the cyst. Stomach and left kidney were displaced towards the right side. There were adhesions with the tail of the pancreas and also dense inseparable adhesions with the diaphragm. Firstly, reduction of cyst with intraoperative drainage of approximately 3 litres of straw coloured fluid was carried out. It was followed by complete splenectomy due to the large size of the cyst, cyst location and presence of adhesions with adjacent structures. Histologically it was a unilocular cyst with dense fibrous cyst wall lined by flattened epithelium. Therefore the histopathological diagnosis was primary splenic (epithelial) cyst.

Post-operative clinical course was satisfactory and at follow up the patient was asymptomatic and was started on lifelong penicillin prophylaxis.

Correspondence: Dr. Prageeth Lambiyas
E-mail: prageeth.lambiyas11@gmail.com

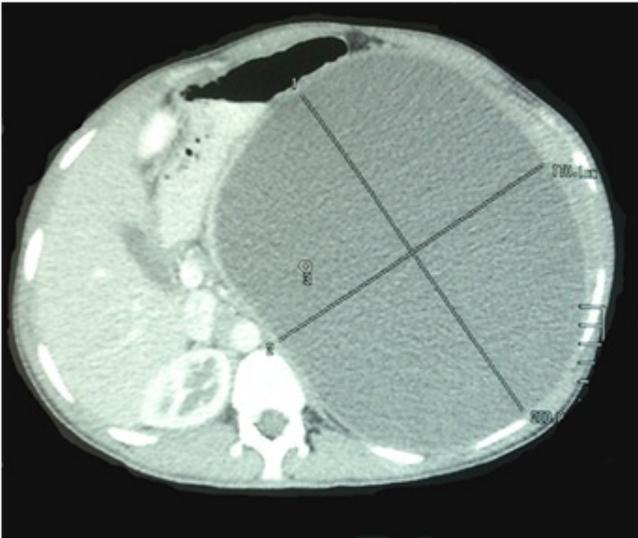


Figure 1. CECT abdomen showing a large cyst compressing the surrounding structure.

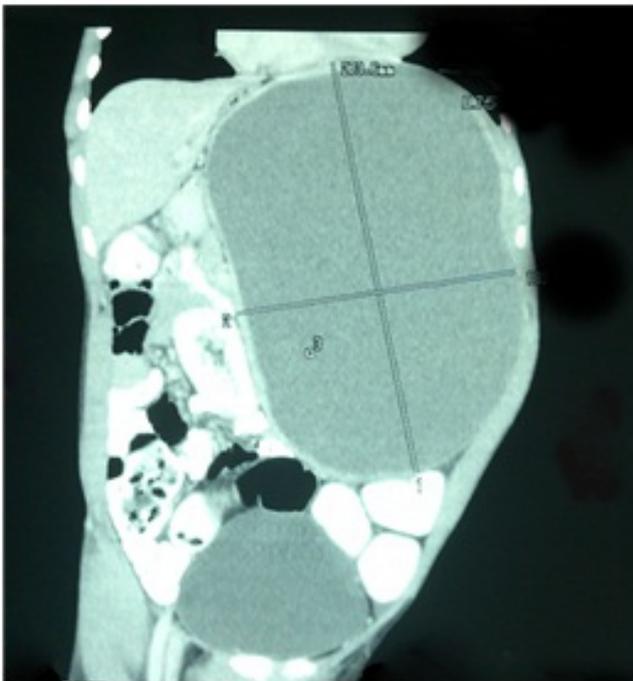


Figure 2. Sagittal section of the CECT abdomen



Figure 3. Intraoperative picture of the cyst

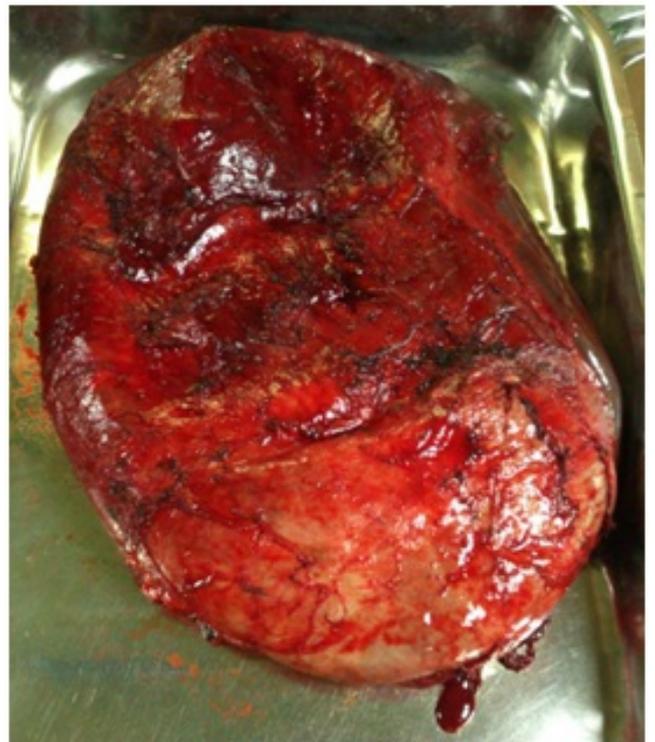


Figure 4. Specimen of the aspirated cyst removed from the patient



Figure 5. Interior appearance of the cyst

Discussion

The spleen plays an important role in haemopoiesis, immune function and protection against infections and malignancies. Splenic cysts are very rare. They are classified as primary (true) or secondary (pseudo/false) cyst. Primary cysts have a cellular lining in the cyst wall and are parasitic or non-parasitic in origin. Non parasitic type primary cysts are further classified as congenital or neoplastic. Congenital splenic cysts are also called epidermoid or epithelial cysts. They are uncommon and account for only 10% of all splenic cysts. 80% of epithelial splenic cysts occur in patients aged under 20 year but can occur in children and infants [4,5].

Secondary cysts without cellular lining usually occur following blunt trauma to the upper abdomen and is considered responsible for 75% of secondary cysts [6].

Both types of splenic cysts usually do not produce any specific symptoms until they reach a significant size and may remain asymptomatic in 30–60% of patients [7]. Splenic cysts may present with localized or referred pain, splenomegaly, abdominal distension and symptoms and signs relating to compression of nearby structures. The latter include early satiety, dysphagia, nausea, vomiting, atelectasis and left lower lobe pneumonia [8]. Cysts may also be an incidental finding on abdominal imaging for another purpose.

A comprehensive differential diagnosis includes cystic lesions of adjacent structures (eg: pancreas, liver, omentum), intrasplenic aneurysms, benign & malignant splenic tumours, pyogenic splenic abscesses and rare parasitic echinococcal disease which is more common in Africa and Central America [9].

Physical examination is usually normal apart from the abdominal distension with abdominal mass. Routine haematological and biochemical investigations are also within normal range. Tumour markers such as CEA and CA19.9 levels are usually elevated in the true cyst and rarely high in pseudo cysts due to the absence of epithelial lining [10,11]. But CA 19.9 and CEA levels were in the normal range in our case.

Indications for operative intervention of the splenic cysts include symptomatic splenic cysts and cysts with a diameter >5 cm because of increased risk of complications [12]. Surgical intervention aims to eradicate the cyst and prevent recurrence. Operative methods include both open and laparoscopic techniques. Traditionally splenic cysts have been treated with the open complete splenectomy. Currently the trend is to more conservative surgery preserving the spleen with the demonstration of life long risk of overwhelming post splenectomy sepsis (OPSI). Post splenectomy patient has 5% life time risk of developing OPSI which carries a mortality rate of 38-69% [13].

Options available for the splenic preservation include total cystectomy, marsupialization, cyst decapsulation (unroofing) or partial splenectomy; accessed either by open laparotomy or laparoscopy. Other conservative

methods include sclerosis or drainage using radiological guidance. The incidence of recurrence of these methods has been reported high as 100% [3]. Partial splenectomy preserves more than 25% splenic parenchyma which is the minimal splenic tissue to preserve immunological protection without increasing the risk of recurrence [14].

In unroofing/ partial splenic cystectomy, the cyst wall should be resected as much as possible to prevent recurrence of the cyst. Marsupialization is another conservative surgery which reduces the duration of surgery and carries no risk of recurrence.

But it is difficult to perform any type of conservative surgery if the cyst is very large; if it is completely covered by the splenic parenchyma (intrasplenic cyst); if it is located in the hilum of the spleen, if there are multiple cysts (polycystic cases) or dense vesicular adhesions to adjacent structures; in these situations, a complete splenectomy should be performed using open or laparoscopic approach [15]

In our case it was impossible to manage with percutaneous drainage and sclerosis technique due to the large size of the cyst. We had to treat a very large cyst located in the splenic hilum with dense inflammatory adhesions around the spleen, whereas splenic parenchyma consisted of rim of tissue pushed to periphery making any possibility of splenic preservation surgery highly unlikely. Therefore the patient was treated with open complete splenectomy.

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Key Points:

- Splenic cysts are a rare phenomenon now increasingly identified due to increased imaging
- Symptomatic and large cysts are an indication for surgery
- Splenic preservation surgery is the treatment of choice when possible