UNUSUAL LONG TERM COMPLICATIONS OF A SPLENIC CYST


Splenic cysts are relatively uncommon, and are usually asymptomatic. They are benign, typically treated conservatively and followed up with ultrasound examination, with few reported complications. We report a case of a simple splenic cyst that was followed up on imaging over a seven-year period. During that time, the cyst gradually enlarged from 5 cm to 12 cm in diameter, however the patient remained asymptomatic. After seven years, the patient was admitted with abdominal pain and a pelvic mass. The spleen was located within the pelvis, which was felt to be due to the weight of the cyst which caused the spleen to migrate out of its normal position. This case illustrates an extremely unusual complication, and suggests that while most splenic cysts may be managed conservatively, enlarging cysts may be prone to gravitational effects and prophylactic treatment should be considered.

Key-words: Spleen, cysts – Spleen, US – Spleen, CT.

Case report

An 18-year-old female was referred by her primary physician to our institution for the assessment of right iliac fossa pain over the previous three days. She had no other abdominal symptoms and her clinical examination was normal apart from some focal tenderness in the right iliac fossa. Laboratory tests were all normal. An ultrasound of the abdomen and pelvis was performed to exclude conditions such as ovarian cystic disease or acute appendicitis. This confirmed that the uterus, ovaries and other pelvic structures were normal. There were no radiologic features of appendicitis. However, it was incidentally noted that there was a 5 cm cyst within the lower pole of the spleen (Fig. 1). The spleen otherwise appeared normal and no other abnormalities were seen. The patient then underwent a non-contrast computed tomography (CT) to evaluate for renal stone disease or occult appendicitis as the cause of her pain. No evidence of renal stone disease or appendicitis was seen. The simple cyst was again seen within a normally positioned spleen (Fig. 2). The patient was managed conservatively and her symptoms resolved. A diagnosis of non-specific abdominal pain was made as well as an incidental finding of a splenic cyst. As the cyst was asymptomatic, it was decided to manage it conservatively by observation with yearly follow up splenic ultrasounds.

Over the subsequent seven years, the patient had annual ultrasounds of the spleen, which demonstrated a gradual increase in size of the splenic cyst from 5 to 12 cm. The cyst appeared simple at all times and the spleen remained in its normal anatomical position. Despite the cyst growing in size over time, the patient remained asymptomatic and it was therefore decided to continue to manage the patient conservatively.

After a seven-year period of follow up, the patient was admitted with new onset acute abdominal pain. A contrast enhanced CT of the abdomen and pelvis was performed. This showed absence of the spleen in its normal position in the left upper quadrant. There however were tortuous varices within the splenic bed (Fig. 3A). These vessels could be traced both to the superior mesenteric vein medially and inferiorly to where the spleen was seen lying within the lower abdomen (Fig. 3B). In the spleen’s dependent portion within the pelvis, the large splenic cyst was identified (Fig. 3C). An ultrasound of the abdomen confirmed the cyst to be simple in nature and with no solid elements and a few simple peripheral septations. An MRI demonstrated the signal of the spleen to be normal with no evidence of infarction (Fig. 4). The splenic cyst appeared of uniform high signal on T2 weighted images suggesting its simple nature. In view of the patient’s symptoms, splenectomy was planned for the following day. Prior to surgery, the patient received meningococcal, pneumococcal and Haemophilus vaccinations. At surgery a very mobile spleen was identified with a large cystic lesion at its lower pole. The spleen was removed and the vessels were divided and tied. Histology showed a 12 cm true epithelial (epidermoid cyst) of the spleen with no evidence of malignancy (Fig. 5). The spleen otherwise appeared unremarkable and the patient had an uncomplicated postoperative course and was discharged home four days later.
Discussion

True cysts of the spleen are relatively rare. They are most commonly discovered within the second and third decades of life. By pathological classification, the presence of a cellular cyst lining, defines a true cyst (1). True cysts represent about one-fourth of nonparasitic benign cysts. False cysts, which have fibrous tissue lining, comprise the remainder. These false cysts may result from trauma, haemorrhage or infarction. Some cysts may also occur secondary to parasitic infestation. Robbins reported a series of 42,327 autopsies over a 25-year period, which revealed only 32 patients with diagnosis of true splenic cyst (2, 3). The origin of splenic epithelial cysts is thought to be congenital although this is controversial. Although no epithelium is present histogenetically within the normal spleen, splenic epidermoid cyst is thought to be a lesion arising from embryonic inclusion of aberrant ectodermal and entodermal epithelium in the developing spleen (4).

Clinically, most patients with splenic cysts are asymptomatic (5). When pain occurs, it is felt to be secondary to the mass effect of the cyst, which compresses the adjacent visceral surface of the spleen. Because of the slow growing rate of epidermoid cysts, symptoms are often vague when present. Symptoms include epigastric fullness and a dull pain in the left upper quadrant. Physical examination is usually negative. Occasionally the spleen can be tender leading to pain on palpation in the left upper quadrant. When the pain is significant, left hemi diaphragmatic movement may be affected leading to lower lobe atelectasis or pneumonia (4). Laboratory tests are usually normal.

The initial diagnosis of splenic cysts is predominantly radiological. Ultrasound typically shows an echo free cystic mass within the spleen with well-defined walls. Rarely calcification may be seen within the walls of the cyst. CT and MRI as in the case we have presented are excellent modalities in demonstrating splenic cysts. They are generally utilised when the diagnosis is in doubt or when complications are suspected.

Splenic cysts are generally incidentally discovered, as they are usually asymptomatic. Although splenic cysts are benign and may spontaneously resolve, some authors suggest that cysts greater than 5 cm should be treated to avoid potential complications (6). The main potential complication of splenic cysts is rupture. This may result in peritonitis or hemorrhage. Other complications include infection and abscess formation (6).

Splenectomy has in the past been the surgical treatment for epidermoid cysts (5, 6). However serious complications can result from this procedure and therefore more conservative measures such as partial splenectomy and laparoscopic cyst decapsulation are now favoured where possible (6).

In the case we have described here, the patient was asymptomatic with an epidermoid cyst, which appeared simple on imaging. The case and options were discussed in detail with the patient and it was decided to manage her conservatively. The risk of rupture is higher in patients who participate in high impact sports. However the patient reported a sedentary lifestyle and this contributed for the decision to manage her conservatively. In retrospect the fact that the cyst had enlarged significantly over her observation period may have been
an indication for surgery. If done at an earlier stage, splenic sparing surgery may have been an option. Due to the large size of the cyst at presentation, splenectomy was considered the safer option.

Migration of the spleen from its normal position within the left upper quadrant is a known phenomenon and often referred to as a wandering spleen. The spleen is normally situated in the left upper quadrant of the abdomen and is held in position by the suspensory ligaments of the spleen, namely the lienorenal and the gastro-splenic ligaments with the phrenico-colic ligament providing further stability.

A wandering spleen is a rare entity that is poorly described in the radiological literature. Its reported incidence is less than 0.2% (7). A wandering spleen occurs when the spleen migrates from its usual position in the left upper quadrant and lies in the left flank, central abdomen or pelvis. Patients may present with a painful mass or an acute abdomen or in some cases they may be asymptomatic (8). In this case, there were varices present as result of the gravitational effect on the splenic vein, when the spleen migrates inferiorly and the length of the splenic vein is therefore increased.

A wandering spleen can be congenital or acquired. In the congenital case there is abnormal development of the dorsal mesogastrium causing defective attachment of the dorsal mesentery. The acquired case is due to abdominal wall laxity or the hormonal effects of pregnancy, which explains why wandering spleen usually occurs in females between the ages of 20-40 years of age (9, 10). Rarely a wandering spleen may be due to an associate mass (11-13) as in the case we have described. CT is the currently the imaging modality of choice in making the diagnosis (13).

**Conclusion**

While splenic cysts are usually asymptomatic and discovered incidentally, they may be prone to complications such as rupture when large. We report an extremely unusual complication of an enlarging splenic cyst that occurred after 7 years of follow up, where the patient presented acutely with a displaced spleen. The displaced spleen was felt to be secondary to gravitational effects by the large cyst. Urgent splenectomy was performed. The case suggests that while small cysts of the spleen may be managed conservatively, the presence of a large or enlarging cyst should make early splenic sparing surgery a consideration.

**References**