Uncommon splenic cysts in paediatric patients: A case series

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SUMMARY

Splenic cysts are uncommon and classified into parasitic and non-parasitic origins. Non-parasitic cysts are further categorised into primary and secondary forms; primary cysts develop congenitally and progress into adulthood and secondary cysts result from factors such as abdominal trauma, infection or ischemia. This case series presents three instances of splenic cysts in children. The first case involves a splenic epidermoid cyst, the second a pseudocyst and the third a splenic epithelial cyst. All patients exhibited an abdominal lump in the left quadrant that increased in size over time, without additional symptoms. The third patient had a history of abdominal blunt trauma a year prior to symptom onset. Treatment approaches varied: the first and third patients underwent total splenectomy, while the second patient underwent aspiration drainage with frozen section analysis and partial splenectomy. All patients, first, second and third, were discharged 6, 3 and 5 days postoperatively, respectively, without complications. Splenic epithelial cyst (SEC) emerged as the predominant primary non-parasitic splenic cyst type, with an unclear pathogenesis. Typically asymptomatic, splenic cysts are commonly detected incidentally during imaging or exploratory laparotomy. Histopathology stands as the gold standard diagnostic method for splenic cysts. Although rare, paediatric splenic cysts should be considered in cases of abdominal trauma. Imaging serves a vital role in diagnosis, guiding decisions between conservative or surgical interventions based on cyst size, symptoms and associated complications.

INTRODUCTION

Splenic cysts are uncommon in children, especially in the youngest age group, while their prevalence is rising as a result of the extensive use of abdominal imaging and nonoperative treatment for splenic injuries.[1] They are traditionally classified as parasitic and non-parasitic origins.[2] Non-parasitic cysts are further categorised into primary and secondary forms, where primary cysts develop congenitally and progress into adulthood, while secondary cysts result from factors such as abdominal trauma, infection or ischaemia.[3] The differential diagnosis is challenging since various spleen-related non-SC diseases, such as inflammatory pseudotumor of the spleen, splenic hamartomas, and splenic abscesses, should be taken into account.[2,3] In this case report, we present three cases of paediatric splenic cysts with different type of cysts requiring splenectomy.

CASE PRESENTATION

Case 1

A 14-year-old male adolescent presented with a lump in the upper left quadrant of his abdomen, accompanied by increasing pain. Despite taking pain medication, the discomfort persisted. Subsequent abdominal ultrasound revealed a uniform hypoechoic mass within the abdominal region. A cystic lesion resembling a splenic epidermoid cyst was identified through an abdominal computed tomography (CT) scan (Figure1). The preliminary diagnosis indicated an intraabdominal mass suspected to be a splenic epidermoid cyst. An exploratory procedure was conducted, uncovering the cystic mass located in the upper-back portion of the spleen. A complete removal of the spleen was performed (Figure 2). Upon histopathological examination, the findings pointed towards an epidermoid cyst in the spleen with cholesterol ester granuloma. The patient's postoperative condition was favourable, and he was discharged after six days.

Case 2

A 14-year-old male adolescent came to the medical facility with a lump in his upper left abdomen, roughly the size of an apple. He didn't have any other complaints over the past 3 years before seeking medical attention. About a year prior to his current complaint, he had experienced blunt trauma to his abdomen. An abdominal ultrasound was performed, revealing a complex cystic mass near the tail of the pancreas that was compressing his left kidney. Subsequent abdominal CT scan indicated a cyst in his spleen that could be an epidermoid cyst, splenic abscess, lymphangioma, or pseudocyst (Figure 1). The provisional diagnosis was an intraabdominal mass suspected to be a splenic epidermoid cyst. The patient underwent a procedure to drain the cyst and a quick analysis during the procedure indicated it was a pseudocyst without signs of malignancy. This was followed by a partial removal of the spleen (Figure 2). The examination of the removed tissue confirmed it was a pseudocyst. The patient was discharged in good condition 3 days after the surgery.

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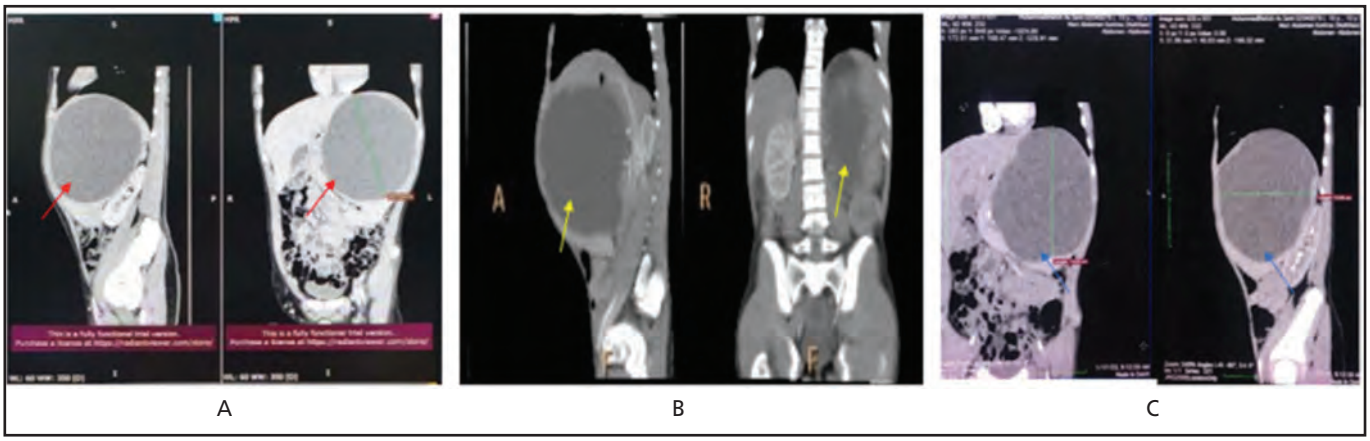

Fig. 1: Abdominal CT scan of case 1, showed splenomegaly with cystic lesion suggested splenic epidermoid cyst with calcification on the wall (A). On abdominal CT scan of case 2, indicated splenic cyst that could be an epidermoid cyst, splenic abscess, lymphangioma or pseudocyst with enlargement to caudomedial (B). On case 3, the cystic mass showed in CT scan exerting pressure on the stomach, duodenum and pancreas (C)

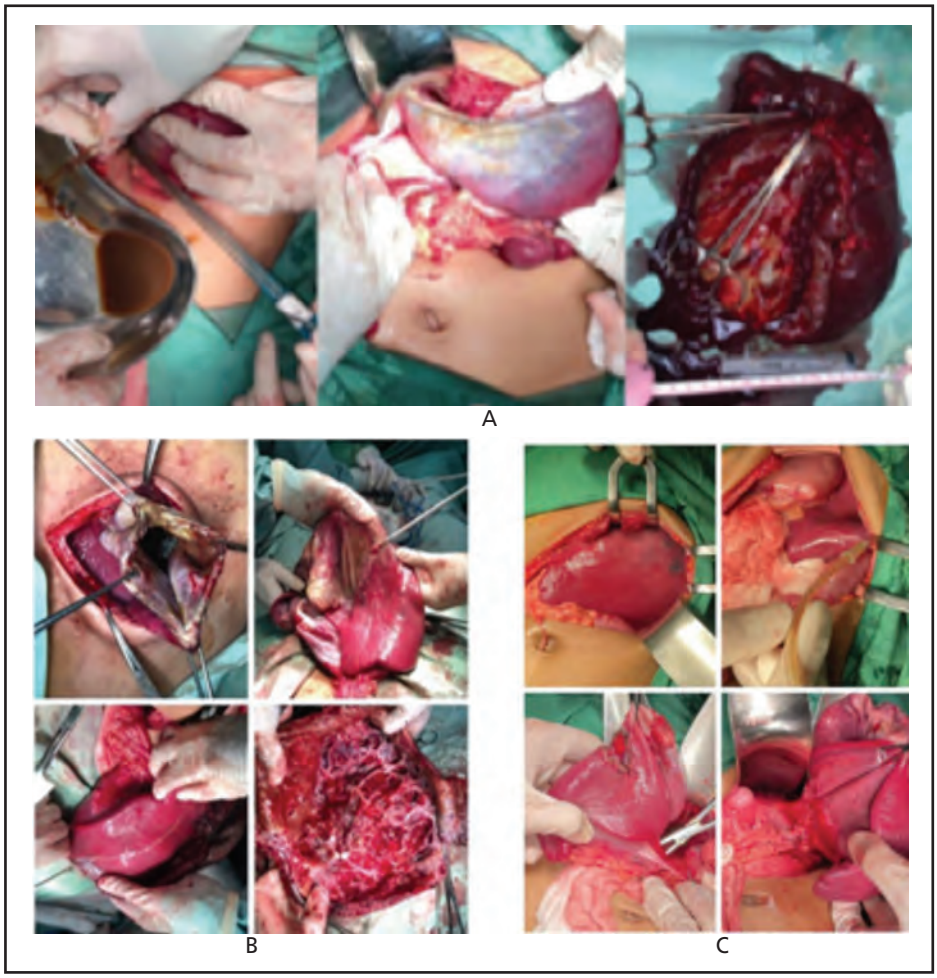

Fig. 2: For case 1, we conduct a laparotomy splenectomy and drainage of the cyst showed brownish fluid around 1300cc (A). The cyst and spleen with separated by clear boundaries in case 2, and we drainage the cyst found serous product around 6000cc (B). In case 3, the cyst wall covers 95% of the spleen with a brownish cystic fluid, so we conducted the total splenectomy (C)

Case 3

A 10-year-old male child arrived with a lump in the upper left quadrant of the abdomen, accompanied by sensations of fullness and loss of appetite. The lump had been progressively growing in size. The patient's medical history included an incident of blunt abdominal trauma a year before the current complaint. An abdominal ultrasound was conducted, revealing a hypoechoic mass situated between the stomach and spleen. Subsequently, an abdominal CT scan displayed a cystic mass within the spleen that was exerting pressure on the stomach, duodenum, and pancreas. This indicated the presence of a splenic epithelial cyst (SEC) (Figure 1). Preliminary diagnosis was an intraabdominal mass suspected to be a splenic epithelial cyst. To address this, a complete removal of the spleen was carried out. The histopathological examination confirmed the presence of a SEC without any signs of malignancy. The patient's recovery was successful, leading to discharge after 5 days following the surgery.

DISCUSSION

We described three cases of paediatric splenic cysts requiring splenectomy. Splenic cysts are rare in children, although their prevalence is increasing due to the use of non-operative abdominal indications of splenic trauma.[1] Splenic cysts are usually seen in the second and third years of life, but they also occur in children and infants. Many factors are discussed about splenic cysts. These congenital diseases go undiagnosed for decades.[1,4] The recurrence rate of this cyst in general is 33% of the total case. Splenic cysts are classified according to Martin's classification, as type I splenic cysts (primary or true cysts), which have a true epithelial lining; and type II splenic cysts (secondary or pseudocysts), which do not have a true epithelial lining and capsule.[5] In the third case, we also examined specific tumour markers to differentiate the cyst from other types such as teratomas, and the results were normal. This finding can distinguish it from other cysts.

Primary splenic cysts are divided into parasitic and non-parasitic cysts. Parasitic cysts are usually seen in endemic areas and are mainly caused by Echinococcus granulosis infestation. Echinococcus granulosis may cause the formation of hydatid cysts, which are sometimes mistaken as epithelial cysts.[4,5] Its can be transmitted through contaminated food, with regions that have high consumption of raw vegetables like salads being at greater risk of infection

Non-parasitic cysts can be divided into congenital cysts and neoplastic cysts. Congenital cysts make up about 10% of all splenic cysts. These cysts can be divided into epidermoid, dermoid and endodermoid cysts. Epidermoid cysts occur due to the inclusion of neighbouring epithelial cells during embryonic development, which then leads to the expansion of the cyst, or from the folding in of the outer layer of cells known as the mesothelium. Dermoid cysts are considered teratoma cysts, containing structures derived from all three germ layers. An endodermoid cyst is a vascular cystic lesion consisting of several ectatic blood vessels. Neoplastic cysts are endothelial origin, include, haemangiomas, lymphangioma, sclerosing angiomatoid nodular transformation (SANT) of the spleen.[3,4]

SEC emerged as the predominant primary non-parasitic splenic cyst type, with unclear pathogenesis. Typically asymptomatic, splenic cysts are commonly detected incidentally during imaging or exploratory laparotomy. Histopathology stands as the gold standard diagnostic method for splenic cysts.[4-6]

Secondary splenic cysts are caused by damage to the spleen or abdominal wall, as well as by splenic infarction or abscess. Distinguishing primary cysts from secondary cysts based on radiographic evidence is challenging due to similarities in features such as calcified or trabeculated cyst walls, peripheral septations, and debris. Splenic cysts caused by injury are not very common, and it is uncertain what the exact frequency of such occurrences might be. It is believed that 75% of secondary splenic cysts, which lead to a splenic hematoma and later develop into an encapsulation with incomplete resorption, are caused by blunt trauma to the upper abdomen. This leads to the accumulation of fluid.[4-6]

Asymptomatic cysts occur in 30 to 60% of patients and are often discovered incidentally, especially those less than 5 cm in diameter. Because cysts grow gradually, they may be observed for years before symptoms appear. Signs and symptoms may include symptoms of pain, swelling, swelling, splenomegaly, or abdominal pressure. The pain may be intermittent or continuous and radiate to the left side, solar plexus, or left shoulder. Splenomegaly causes hypersegregation of the spleen, which can manifest as anaemia, thrombocytopenia and coagulopathy. In our cases, there is no report of any anaemia, thrombocytopenia and coagulopathy before and after surgery.[4,5]

Distinguishing between a true cyst and a pseudocyst using imaging techniques is a challenging task. The cyst can be detected using regular X-ray images if it is significant. A spherical object with a complete and healthy end of the spleen might distinguish between a cyst and an enlarged spleen. Calcification of the arch can also be useful in identifying cysts. On an ultrasound, a splenic cyst typically appears as a clear lesion with smooth edges and shows heightened transmission. If there is flow on Doppler ultrasound, flow lesions such as lymphangiomas and abscesses may be considered. On a CT scan, splenic cysts are observed as uniform, clearly defined lesions and either round or oval in shape. They appear as areas of lower density, not showing significant enhancement or improvement in contrast. The layer surrounding the cyst may become thicker or contain some areas of calcification, and it might sometimes be divided by thin walls. Splenic cysts appear dark on T1-weighted agnetic resonance imaging (MRI) images and bright and consistent on T2-weighted MRI images.[5]

The management of splenic cysts is still controversial. There are no guidelines regarding asymptomatic splenic cysts. Approaches to treat splenic cysts include aspiration and consolidation, internal and external marsupialisation, partial cystectomy (capsule removal), splenectomy and nonsurgical treatment. Symptomatic or complex cysts require

surgical removal.[1-5] Cysts that involve only a portion of the spleen and protruding to the surface can easily be treated by marsupialization, deroofing, or cyst excision. Deep cysts in lower pole that cannot be accessed by this procedure are best treated by partial splenectomy. If the cysts located in the hilar level often requiring complete splenectomy due to bleeding risks. Cyst aspiration as a definitive treatment has been described previously. Agents such as tetracycline or alcohol are injected into the cyst to destroy the cyst lining, but recurrence still occur. Therefore, this procedure is intended only as a temporary management.[7]

We performed splenectomy on three individuals who had splenic cysts. The surgical removal of splenic cysts is typically recommended, due to the risk of infection, rupture from injury, severe bleeding, or the development of abscesses. In the past, the usual approach to deal with splenic cysts involved removing the entire spleen. However, there has been a suggestion to use more cautious methods in order to preserve a greater amount of splenic tissue. In certain cases, total splenectomy may still be recommended, particularly when dealing with polycystic or very large cysts that are mostly concealed within the spleen tissue. This approach is taken to avoid the potential risk of bleeding.[9] Partial splenectomy approach is used to reduce splenic volume (85 to 95%), leaving about 10 to 25% of the normal spleen remaining. The main advantage of partial splenectomy is that it preserves the immune function of the spleen, therefore reduce the risk of post-splenectomy sepsis. The use of laparotomy or laparoscopy can be performed, and generally depends on the skill and preference of the surgeon.[8]

Recovery after total splenectomy is rapid, and most children are discharged from the hospital the day after laparoscopic surgery. Partial splenectomy usually requires the patient to stay in the hospital for 2 to 3 days so that he can be monitored for signs of bleeding. Open surgery requires a slightly longer recovery time to reduce pain, but most children are discharged from the hospital relatively quickly after surgery. To prevent overwhelming post-splenectomy infection (OPSI), antibiotic prophylaxis and immunisation are recommended in the early postoperative period. Vaccinations against Streptococcus pneumoniae, Haemophilus influenzae, and Meningococcus are recommended due to their common association with OPSI. Penicillin is the preferred antibiotic according to most experts, though trimethoprim-sulfamethoxazole can be used as an alternative for those allergic to penicillin. Although there are no definitive guidelines on the duration of prophylaxis, it is generally recommended for at least 2 years following splenectomy. Portal vein thrombosis, occurring in approximately 4.79% of cases, is a relatively rare complication following a splenectomy. Indicators for this condition may include spleen size and thrombocyte counts. Pulmonary hypertension (PH) is another reported complication, with an incidence of about 10%. Although PH is thought to result from thromboembolic involvement of the pulmonary microvasculature through increased thrombus formation, the precise mechanism remains unclear.[10]

CONCLUSION

This study presented three cases of paediatric splenic cysts requiring splenectomy. Despite their rarity in young children, their occurrence is on the rise due to advanced imaging and evolving trauma management. The classification, clinical features, diagnostic challenges, and varied management approaches were explored. From asymptomatic to symptomatic cases, the complexity of paediatric splenic cysts and the nuanced decisions involved in their treatment were highlighted.

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CONSENT STATEMENT

All patients involved in this study provided informed written consent. Their identities have been kept confidential throughout the study.

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