Case Report

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A report of a giant epidermoid splenic cyst

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SUMMARY

Splenic cysts are uncommon lesions and are classified as true or pseudocysts based on the presence of an epithelial lining. True congenital cysts can be epidermoid, dermoid or endodermoid and require surgery when large, sympthomatic or complicated. Ultrasonography and computerised tomography scan are useful for diagnosis and serum test for echinococcosis should be done. Several procedures, open or laparoscopic, have been described, with emphasis on the spleen-preserving surgery, when possible. We present a boy with a giant symptomatic epidermoid cyst in whom total splenectomy by open technique was carried out.

Key words: Children, epidermoid cyst, management, splenic cyst

INTRODUCTION

Splenic cysts are unusual in everyday surgical practice, especially in western countries and less than 1000 cases have been described in the literature.[1] They are classified as true (primary, 25%) lined by epithelium or false cysts (pseudocysts or secondary, 75%) without epithelial or endothelial lining (often secondary to trauma, haemorrhage or infarction).[2] True cysts can be further subdivided into parasitic (caused by *Echinococcus granulosus*) and nonparasitic[3] according to their aetiology. Nonparasitic cysts are neoplastic (lymphangioma and haemangioma) or congenital which are classified as epidermoid (90%), dermoid and endodermoid depending on the type of lining[1,2] [Table 1]. Usually, congenital cysts are benign, sporadic, with a slight female preponderance, present at younger age and their pathogenesis is unclear. Most of the cysts are asymptomatic presenting as an incidental finding during the abdominal ultrasonography (USG). Some patients present symptoms related to complications or bulky size. In fact, cysts larger than 5 cm are susceptible to haemorrhage, rupture and infection and surgery is recommended for congenital cysts >4 cm.[1] USG and computerised tomography (CT) are useful investigations but diagnosis of an epidermoid cyst is only made by the pathologist. Controversies exist regarding optimal surgical management (aspiration, marsupialisation, cystectomy, decapsulation, partial splenectomy, splenectomy). In fact, the increased risk of overwhelming post-splenectomy infection in children[4] has prompted organ-saving procedures. We report a case of a 9-year-old boy with a giant epidermoid splenic cyst with discussion on surgical strategies.

Table 1: Splenic cysts classifications according to Fowler, Martin and Morgenstern

Fowler	Martin	Morgenstern
Primary with epithelium	Primary (true cyst)	Congenital
Congenital	Parasitic	Mesothelial, transitional, epidermoid-epithelial tissue
Serious		
Transitional		
Traumatic	Nonparasitic	Without lining
	Congenital	
	Neoplastic	
Inflammatory		Neoplastic
Neoplastic		Endothelial
Epidermoid		Limphangioma
Dermoid		Haemangioma
Lymphangioma		
Haemangioma		
Secondary without epithelium	Secondary (pseudocyst)	Cystic primary tumours or metastasis
Traumatic		Traumatic
Degenerative		Degenerative (post-infarction)
Inflammatory		

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CASE REPORT

The present case report is about a 9-year-old boy who was admitted to our Unit because of abdominal pain localised in the left upper quadrant that had started a day before. The patient's medical history was unremarkable, including previous abdominal trauma. The physical examination revealed only a distended and asymmetric abdomen with a palpable mass in the left hypochondrium. Routine blood tests were within normal ranges. Chest and abdominal X-rays showed the elevation of the left hemidiaphragm and asymmetric intestinal gas distribution. The abdominal USG revealed a giant, round hypoechoic and multilocular cystic lesion (maximum diameter of the cyst was 20 cm) with internal echoes without Doppler flow. The abdominal CT scan demonstrated a well-limited, large multilocular (20 cm × 13 cm × 21 cm), non-enhancing and hypoattenuated cystic lesion occupying all the splenic parenchyma causing medial displacement of the stomach, pancreas and left kidney [Figure 1]. The cyst was septed with smooth wall and fluid content. These findings oriented for the diagnostic hypothesis of hydatidosis or congenital cyst of the spleen. However, serology for *E. granulosus* was negative so that surgery was planned. After pre-treatment with meningococcal, pneumococcal and haemophilus influenza vaccines, antibiotic and steroid prophylaxis, the child underwent open splenectomy. Splenectomy was indicated due to giant size, the location at hilum and the residual atrophic splenic tissue [Figure 2]; at surgery, we observed that most of splenic blood flow was originating from a branch (right gastroepyploic) of gastroduodenal artery while the splenic artery was completely hypoplasic. The post-operative course was uneventful and he was discharged 5 days later. Pathological examination showed an epidermoid congenital cyst with fibrous tissue covered by stratified squamous epithelium [Figure 3] without evidence of epithelial atypia or malignancy. At immunostaining, the epithelium was positive for cytokeratines AE1 and AE8 [Figure 4]. The

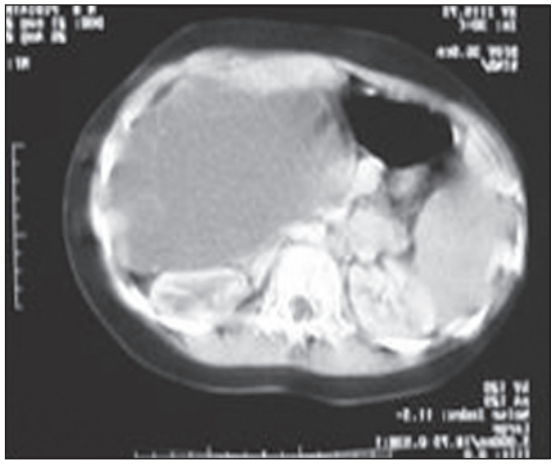


Figure 1: Computerized tomography scans showing the giant splenic cyst

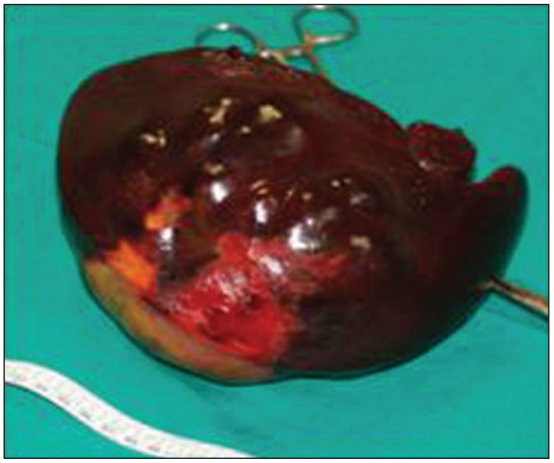


Figure 2: Surgical specimen: Spleen with epidermoid cyst

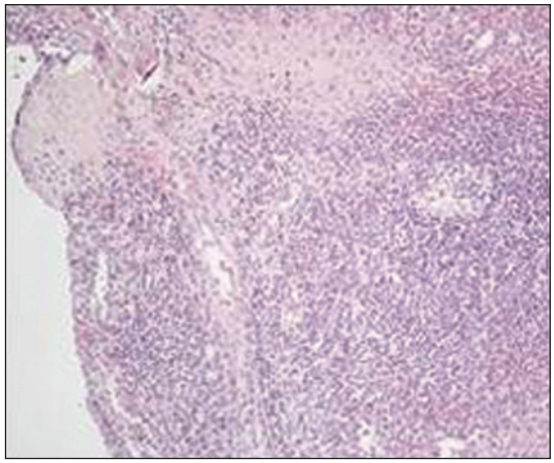


Figure 3: Epidermoid cyst with squamous epithelium (EE ×40/×100)

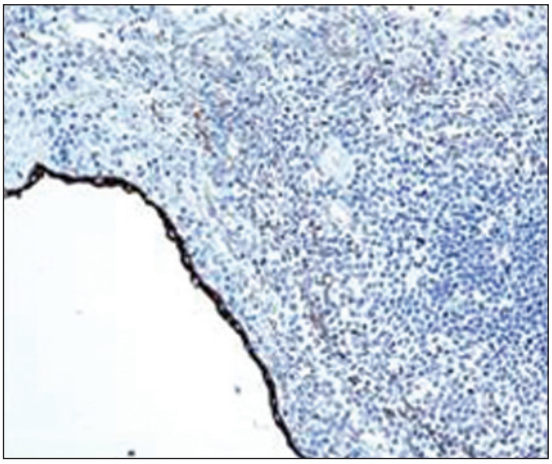


Figure 4: Positivity at immunostaining for cytokeratines AE1 and AE8 (×200)

fluid was turbid and yellow with some cholesterol crystals and lipids. At 12 months follow-up, the child is doing well with normal blood or immunological tests.

DISCUSSION

The incidence of splenic cysts is low and congenital ones are very uncommon.[5] These are usually large, solitary and sporadic (few familial cases have been reported without identifying genetic predisposing factors).[6,7] The physiopathology is still controversial especially, about their origin from developmental displacement of epithelial tissue, with subsequent metaplasia, or whether they arise from the peritoneal mesothelium. The types of epithelial tissues hypothesized are germinal cells from gonads,[8] the dorsal mesogastrium[9] and the Wolffian duct.[10] Bürrig[11] and Ough *et al*.[12] have reported that epithelial cysts originate from invaginations of capsular peritoneal mesothelium or collections of peritoneal mesothelial cells trapped into splenic sulci, which subsequently undergo to squamous metaplasia (possibly as consequences of chronic inflammation). Furthermore, Authors suggested that epithelial cysts have a teratomatous derivation[13] while Others[14] considered splenic epithelial cysts the result of secondary fluid collection after injury or spontaneous intrasplenic bleeding. In addition, communication between the pancreatic duct system and the intrapancreatic accessory spleen might lead to cyst growth.[15] The clinical manifestation differs according to size and the symptoms are non-specific and are related to compression of the adjacent organs. Malignant degeneration of primary splenic cyst is rare. Serum carbohydrate antigen 19-9 and carcinoembryonic antigen have been reported to be elevated in serum and cyst fluid[16] but these antigens were not investigated in our patient. USG and CT are the most frequently used diagnostic methods helpful in determining whether the cyst is multi- or unilocular, the location of the cyst and relationship to the surrounding structures.[17] USG is a sensitive, cheap and non-invasive tool in the diagnosis and epidermoid cyst appears as a round anechoic area with thin wall. Septations and calcifications may be seen. On CT, cysts are spherical without rim enhancement. The conventional treatment of splenic symptomatic, large or complicated cysts[18] has been total splenectomy, open or laparoscopic. In recent years, a spleen-preserving surgical approach is recommended since it is well-known that the spleen plays an important role in normal homeostasis (regulation of the circulating blood volume, haematopoiesis, immunity and protection against infections and malignancies). In fact, the risk of post-splenectomy sepsis is about 4%, with a mortality rate of 1.5%.[19] However, conservative approaches such as percutaneous aspiration or sclerosis (by using tetracycline or alcohol) are associated with high recurrence rate and potential risk of bleeding and infection.[20,21] In addition, dense inflammatory response around the spleen makes subsequent surgery difficult. Today the suggested treatment options are partial splenectomy (preserving >25%), cystectomy, decapsulation and marsupialization, with open or laparoscopic approach. Partial splenectomy is the best known spleen-preserving procedure and is based on the segmentation of the splenic vascularization. The first successful partial splenectomy for an epidermoid cyst was carried out in 1980[22] but it is a difficult procedure with higher risks of major intraoperative and post-operative blood loss. Cystectomy is the removal of the cyst wall with lining epithelium and a portion of the contiguous splenic parenchyma, but it can be attempted only in superficial cysts that are not close to the ilum.[1,17] Furthermore this technique may be complicated by major intra-operative blood loss. Some Authors have proposed decapsulation of the cyst, leaving the portion of cyst wall contiguous with the splenic parenchyma *in situ*.[23,24] Its advantages include a simpler and more rapidly performed procedure, with less blood loss, simplified haemostasis and preservation of splenic tissue;[25] disadvantage is higher recurrence rate because a portion of the cyst lining is left intact. Marsupialisation is creation of an opening on the cyst wall for internal or external drainage[26] with placation of cyst edges to prevent collapse of the cyst wall and recurrence. Recently, advances in operative techniques, haemostasis with fibrin glue, radiofrequency ablation and stapler techniques have made spleen-preserving procedures safe and feasible.[27-30] However, total splenectomy remains the treatment of choice if the cyst is big enough to involve all the spleen which is reduced to a small and atrophic remnant, as it was in our case, or if the cyst involves the hilum. In addition, complete removal of the cyst is needed since recurrence has been reported after open and laparoscopic surgery.[31] Total splenectomy remains a safe procedure, associated with few complications. Laparoscopic approach has recently gained increasing acceptance in splenic surgery and was first reported in the adult literature.[32] In the paediatric experience it showed good short-term results[33,24] but the largest retrospective studies showed that it is associated with a high recurrence rate.[5,34-36] The significance of splenic cyst recurrence after laparoscopic treatment remains unclear and it ranges from 50% to 88% and may depend on the type of laparoscopic resection performed. Furthermore, laparoscopic surgery provides a minimal

access method to approach small true cysts, but in our patient the cyst was giant (maximum diameter 20 cm) and the splenic parenchyma was completely occupied and substituted by the mass. Despite the pre-operative serology for *E. granulosus* was negative the cyst was multiloculated and the risk of eventual dissemination of cyst fluid could not be excluded, that is why we decided to perform a laparotomy.

CONCLUSION

In summary, the epidermoid cyst is a rare splenic lesion and usually presents as a large cystic mass producing abdominal symptoms. There is no reliable pre-operative diagnostic method to surely identify an epidermoid cyst. The laparoscopic management offers the benefits of minimally invasive surgery (minimal post-operative pain, faster recovery, shorter hospital stay, better aesthetic result) but has an higher recurrence rate, especially in partial splenectomy. The spleen-preserving techniques should be attempted when nonparasitic cysts are relatively small and peripheric. We believe that in our patient, total splenectomy was the safest and the most effective procedure to treat giant epidermoid splenic cyst.

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