

Case report

Giant Epithelial Cysts of the Spleen in Children: Report of Two Cases and Mini Literature Review

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SUMMARY

Introduction. Cystic changes of the spleen are one of the rare clinical entities in the pediatric population. The paper presents a partial splenectomy in children performed by open approach as a safe surgical treatment of giant epithelial splenic cysts.

Case report. Two cases of giant epithelial cysts of the spleen in children are presented. Medical documents and data referring to age, clinical features, findings on ultrasonography and magnetic resonance imaging (MRI), performed surgery, and follow-ups were collected and analyzed retrospectively. Both of the patients complained of abdominal pain; the diagnosis was made on ultrasound and confirmed by MRI. Open cystectomy with partial resection of the spleen was performed in both cases. There were no complications and no recurrence during the follow-up period. The literature review revealed that up to date only seven authors have published papers on this topic, and different treatment methods and results have been reported.

Conclusion. Although very rare, splenic cysts should always be considered in the differential diagnosis of non-specific pain or cystic formations in the upper left quadrant of the abdomen. Taking into account the immunological role of the spleen, tissue preservation surgery should be the main goal. Open partial splenectomy which was performed in both patients was without intra- or postoperative complications and uneventful follow-up, and could be one of the safe management options.

Keywords: epithelial cysts, spleen, children

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INTRODUCTION

Cystic changes of the spleen are one of the rare clinical entities in the pediatric population. Literature data shows an incidence of 0.5–2%, where half of the total number occurs in the age under 15 years (1). Based on the presence of an epithelial lining, all cystic changes of the spleen are divided into true or primary cysts that are characterized by the presence of an epithelial lining and pseudocysts or secondary cysts that do not have an epithelial lining. By further division, primary cysts can be divided into non-parasitic or congenital and parasitic (2). The most common cause of primary cysts of the spleen is human hydatid disease or echinococcosis, commonly seen in Africa and Central America. However, non-parasitic cysts are much less common and usually occur in Europe and North America (3). The exact pathophysiological mechanism of the occurrence of these cysts has not yet been clearly defined and there are several different theories such as mesothelial invagination theory, lymph space theory, and endodermal inclusion theory (4-6). Although diagnosing splenic cysts are not such a

challenge, the management of these cysts could be challenging. Considering the lack of an adequate amount of data, there is still no clearly defined standard for the treatment of these changes in the pediatric population. This study aims to present a partial splenectomy in children performed by open approach as a safe surgical treatment of giant epithelial splenic cysts.

CASE ONE

Medical documents and data referring to age, clinical features, findings on ultrasonography and MRI, performed surgery, and follow-ups were collected and analysed retrospectively.

The first patient was a twelve-year-old girl who presented to the pediatric surgeon due to the existence of a painless lump in the left hypochondriac region. An ultrasound examination was performed and a cystic lesion in the spleen was revealed. A magnetic resonance imaging of the abdomen findings confirmed a large splenic cyst measuring 105 x 76 x 85 mm (Figure 1). Tumor

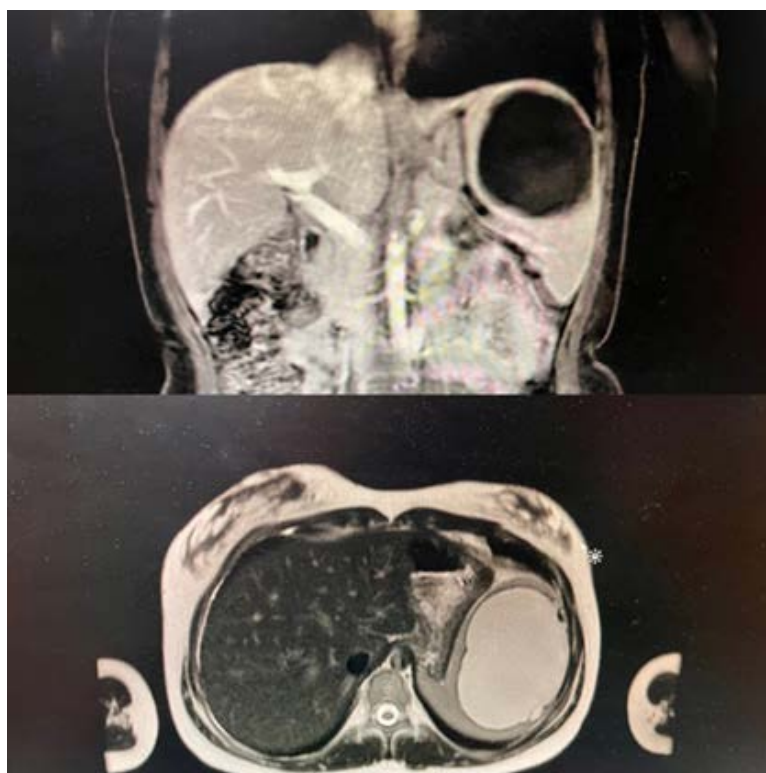


Figure 1. Magnetic resonance imaging – the first patient

markers were increased: CEA (carcinoembryonic antigen) was 8.4, and Ca 19-9 was 152.3. Other laboratory findings were within normal values. Cystic echinococcosis was excluded using a serology ELISA test (enzyme-linked immunosorbent assay). Preoperatively, the patient was given a pneumococcal vaccine in case that splenectomy was needed.

Open cystectomy with partial resection of the spleen was performed. Initially, 450 ml of fluid was evacuated from the cyst. The Ulttacion Harmonic Scalpel (Ethicon) was used for spleen resection. Haemostasis was controlled with SURGICELTMAbsorbable Hemostat (Ethicon) and interrupted sutures with Vycril 3/0. No transfusion was required. There were no postoperative complications, evidence of bleeding, or related readmissions. No long-term antibiotics were given after discharge from

hospital. The final diagnosis was based on the pathological study of operative specimens.

A unilocular, emptied cystic structure was submitted for pathohistological diagnosis. Micro-morphological analysis of the sampled tissue on histological sections revealed a cyst with relatively thick walls. The inner part of the cyst was lined with squamous epithelium. Cellular atypia and pathological mitoses were not observed. The expression of CEA and CA 19-9 was observed by immunohistochemical analysis (Figure 2 and 3).

Regular follow-ups, firstly after a month and afterward every three months for physical and ultrasound examination, and blood cell count in the outpatient department were performed. No cyst recurrence was observed.



Figure 2. *The inner side of the splenic cyst*

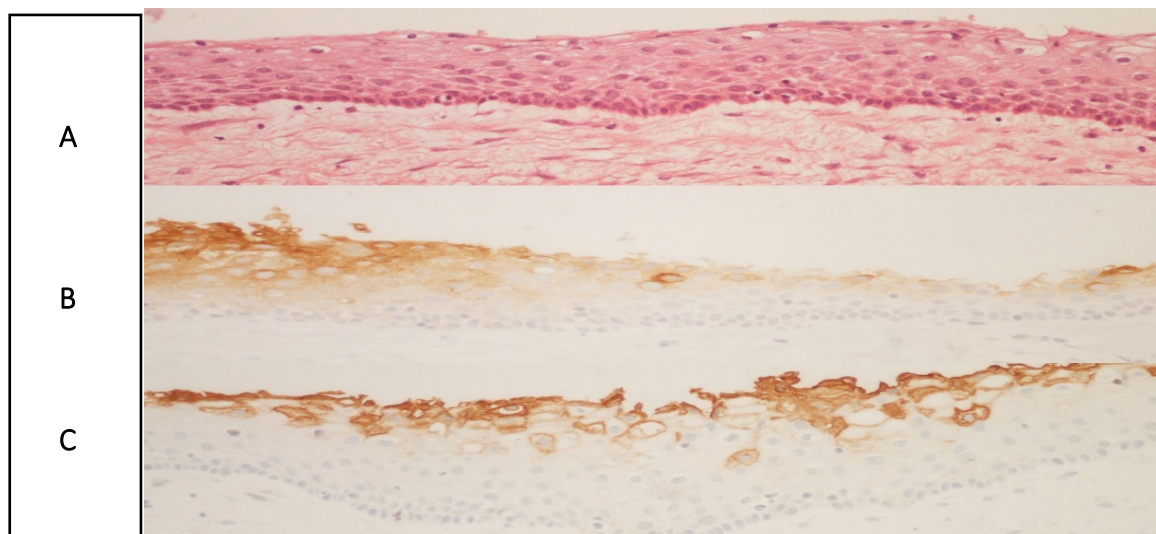


Figure 3. Histopathological and immunohistochemical analysis. A - H&E staining (magnification x 40); B - CEA expression (magnification x 40); C - Ca 19-9 expression (magnification x 40)

CASE TWO

The second patient was a ten-year-old girl who had experienced episodes of abdominal pain a few months before the physical examination. The pain was present periodically, especially when she was physically active. A huge cyst in the spleen was first detected by ultrasound, and later, the patient underwent an abdominal MRI examination when a gigantic splenic cyst was described with dimensions 143 x 114 x 135 mm (Figure 4.) All laboratory findings and tumor markers were in the normal range except Ca 19-9 which was 47.3. Echinococcosis was excluded preoperatively, and immunization against the bacteria *Streptococcus pneumonia* was conducted.

Although the cystic lesion was gigantic, and 850 ml of fluid was evacuated from the cyst at the beginning of the surgical procedure, open cystectomy with partial splenectomy was performed and the lower pole of the spleen was preserved (Figure 5). Resection and haemostasis were done in the same manner as previously described in the first case. There were no complications during the surgery or postoperatively, and there was no need for transfusion also. The diagnosis of the epithelial splenic cyst was confirmed on histopathological examination. There was no evidence of cystic rests or recurrence during the follow-up period.

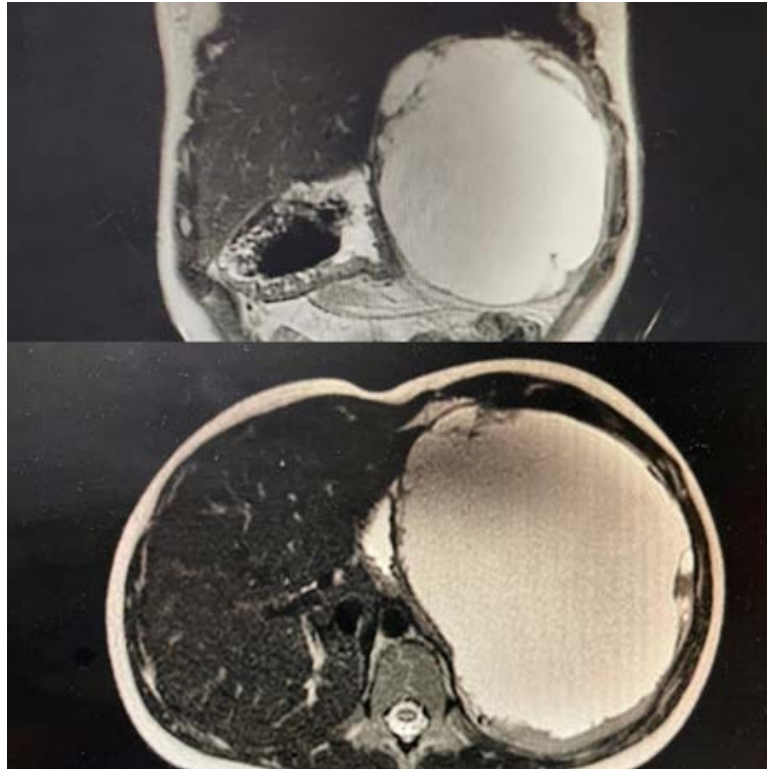


Figure 4. Magnetic resonance imaging – the second patient

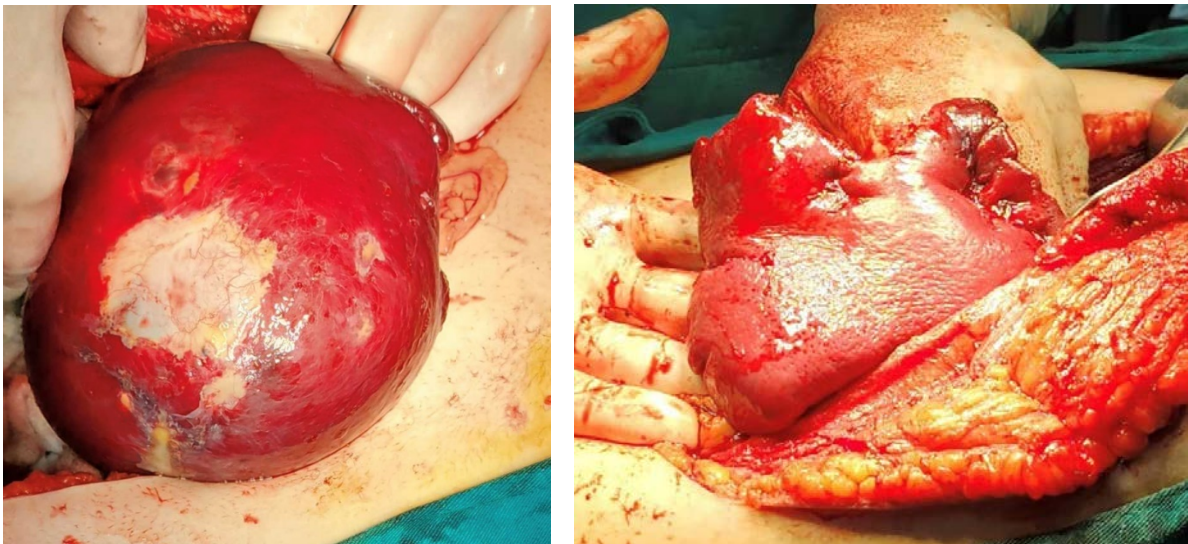


Figure 5. A - Macroscopic appearance of the cyst; B - Resected spleen after cystectomy and partial splenectomy

MINI LITERATURE REVIEW

A mini literature review was conducted on this topic. English and Serbian literature was searched on PubMed, Medline, and Kobson databases for terms: cysts, spleen, children, and surgery. Data were entered in a Microsoft Excel (Microsoft Corporation) spreadsheet containing the author's name, year of publication, number of patients, (age),

types of performed surgery, complications, and if there were rests of the cysts. All papers that matched our inclusion criteria, containing data on children who were treated due to epithelial splenic cysts were included. Patients with parasitic cysts and literature reviews were excluded. Results are given in Table 1.

Table 1. Authors, year of publication, patients, and type of surgery are represented in the table

Author	Year of publication	Number of patients	Gender (male and female)	Age (years)	Open/Laparoscopic surgery	Type of surgery	Other	Complications	Remnant/ Recurrence
Tsakayannis De	1995	19	M10/F9	0-17	14 open surgeries	Splenectomy 9 Partial splenectomy 2 Cystectomy 3	5 cysts were found on autopsy	/	/
Gezer HO	2015	22	M13/F9	0-14	9 open, 1 laparoscopic surgeries	Partial splenectomy 2 Cystectomy 5	12 ultrasound Follow-up	/	/
Zvizdic Z	2012	1	M0/F 1	10	1 open surgeries	Cystectomy 1	/	/	/
Hassoun J	2007	42	M19/F23	0-18	2 open, 6 laparoscopic surgeries	Splenectomy 3 Partial splenectomy 2 Cystectomy 3	32 ultrasound follow up (in 4 patients aspiration was performed)	/	/
Pang W	2009	10	M8/F2	5-15	5 open surgeries	Splenectomy 4 Partial splenectomy 1	Other 5 patients were in angioma group	/	/
Czauderna P	2006	50	M26/F 24	1-17	26 open, 24 laparoscopic surgeries	Partial splenectomy 26 Cystectomy 9 Derroofing 9	/	2 massive bleeding	Eight remnants of the cyst after laparoscopy
Kimber C	1998	6	M2/F4	8-16	6 open surgeries	Splenectomy 1 Partial splenectomy 5	/	/	/
Marjanovic Z	2008	5	M2/F3	13-24	5 open surgeries	Splenectomy 1 Partial splenectomy 4	/	/	/.. 0

DISCUSSION

Splenic cysts in children are not common and clinical presentation and symptomatology are mostly non-specific and depend on the size of the cyst. Smaller cysts are usually asymptomatic, while larger cysts can be presented by a palpable abdominal mass, abdominal pain, vomiting, coughing, or some complications in the form of infection, rupture, and subsequent hemorrhage. Continuous or inconsistent abdominal pain, or radiating left shoulder pain is a consequence of capsule distension or the consequence of dislocation of other abdominal organs (7–9). The increase in the volume of the cyst may be due to the proliferation of the epithelial lining and its secretion or bleeding (10), or it may be due to a difference between the osmotic pressure of the surrounding tissue and the inside of the cyst (11). A palpable mass was present in the first and periodical abdominal pain in the second patient.

The vast majority of splenic cysts are detected accidentally by different imaging methods, most often after abdominal trauma. Diagnosis is commonly made by ultrasound examination. Multislice spiral computed tomography (MSCT) and MRI can give concrete data about the morphology of the cyst, whether it is uni- or multilocular, information about the relationship with the surrounding structures, the content inside the cyst, the existence of pathological communication, the localization of the spleen, so the surgeon can preoperatively make the optimal operative approach (12). Both of our patients underwent an ultrasound examination first. The diagnosis of splenic cysts was confirmed later on MRI. Regarding biomarkers, serum carbohydrate antigen (CA 19-9) can be elevated, and it should be measured preoperatively and postoperatively to exclude recurrence. Also, carcinoembryonic antigen (CEA) with CA 19-9 should be used for differentiating benign cystic lesions from mucinous cystic neoplasm of the pancreas (13). These markers were elevated in both of our patients.

The management of splenic cysts in children includes several options such as observation, aspiration with or without sclerotherapy, deroofting, cystectomy, partial or total splenectomy, and open or laparoscopic approach. Some authors recommend regular follow-up with control ultrasound examinations of cysts smaller than 5 cm as spontaneous regression is described, however, cysts larger than 5 cm should be treated surgically (14, 15). Another

type of non-surgical invasive treatment is percutaneous aspiration with or without sclerotherapy. The main problem of this type of treatment is recurrence (16, 17). The literature is scarce about the type of treatment in the pediatric population, as it is presented in Table 1. Up today, only 145 pediatric splenic cyst cases have been published. Nowadays, taking into account the immune role of the spleen, tissue preservation surgery is the main goal. Total splenectomy is performed if splenic cysts are located in the hilum of the spleen, in the presence of very large or multiple cysts, and in case of excessive intraoperative bleeding (18). The minimal percentage of tissue required for the normal immunological function of the spleen is 25% (19), therefore, recommended surgical options include partial splenectomy, total cystectomy, deroofting, or marsupialization of cysts. Open partial splenectomy is a relatively safe procedure, without recurrence after this type of surgical treatment (20, 21). Tsakayannis et al. published that all of their patients treated before 1971 underwent splenectomy, and patients treated afterward had hemisplenectomy or cystectomy (22). Also, Pang et al. reported that the majority of their patients had total splenectomy, and one had partial splenectomy (23). Calisti et al. reported five children treated with open total cystectomy, which means peeling the cystic wall off the splenic tissue. After a follow-up period of 6 months to 10 years, they did not show recurrence and had no major complications except in one case (24). A case similar to our second patient in age, gender, cyst dimensions, and treatment was reported by Zvizdic and Karavdic (25). In the last decades, laparoscopy has become the method of choice for many surgeons. The main advantages of laparoscopy include shorter hospital stays, faster recovery, less postoperative pain, and better cosmetic outcomes, but there are also longer operative times increasing operative costs, and high recurrence rates as disadvantages of this method. Hassoun et al. reported recurrence after laparoscopic cystectomy in one patient (26); also, Sinha et al. reported 50% of recurrence after cystectomy. They believed that the high rate of recurrence is connected with the laparoscopic approach (27). Nowak et al. described laparoscopic technique for decapsulation, which includes suctioning out the cyst, opening and excising the cystic wall with LigaSure or Harmonic Scalpel device which is more safe as there is less blood loss compared to excising of spleen parenchyma. One should bare in mind that

any remaining fragment of cyst wall could lead to recurrence of the cyst (28). Laparoscopic fenestration is also a safe method for the treatment of superficially located cysts. This method involves the resection of the cyst wall and the creation of permanent communication between the cystic cavity and the peritoneum, however, there is a high rate of recurrence after this technique. Czauderna et al. reported recurrence in 7 of 9 patients treated with laparoscopic fenestration or deroofing (29).

CONCLUSION

Although very rare, splenic cysts should always be considered in the differential diagnosis of

non-specific pain or cystic formations in the upper left quadrant of the abdomen. After the diagnosis, further therapeutic treatment depends on the size of the cyst, and therefore, cysts of smaller dimensions (< 5 cm) should be monitored with regular ultrasound examinations, while for larger cysts there are several different therapeutic modalities. Taking into account the immunological role of the spleen, tissue preservation surgery should be the main goal. Open partial splenectomy which was performed in both patients without intra or postoperative complications and uneventful follow-up could be one of the safe management options.

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Gigantske epitelne ciste slezine kod dece: prikaz dvaju slučajeva i mini-pregled literature

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SAŽETAK

Uvod. Cistične promene slezine predstavljaju retku kliničku pojavu u pedijatrijskoj populaciji. Cilj ovog rada bio je da predstavi otvorenu parcijalnu splenektomiju kod dece kao bezbedno hirurško lečenje gigantskih epitelних cista slezine.

Prikaz slučaja. Prikazana su dva slučaja džinovskih epitelnih cista slezine kod dece. Retrospektivno su prikupljeni i analizirani medicinski dokumenti i podaci koji se odnose na godine starosti, kliničke karakteristike, nalaze sa pregleda ultrazvukom i magnetnom rezonancom, urađene operacije i praćenja. Oba bolesnika su se žalila na bolove u trbuhu. Dijagnoza je postavljena ultrazvučnim pregledom i potvrđena snimanjem magnetnom rezonancom. I u jednom i u drugom slučaju urađena je otvorena cistektomija sa delimičnom resekcijom slezine. Nije bilo komplikacija i recidiva za vreme praćenja. Pregled literature ukazao je na to da je samo sedam autora objavilo radove o ovoj temi, kao i da su prikazane različite metode lečenja i rezultati.

Zaključak. Mada su veoma retke, ciste slezine uvek treba uzeti u obzir prilikom diferencijalne dijagnoze nespecifičnog bola ili cističnih formacija u gornjem levom kvadrantu trbuha. S obzirom na imunološku ulogu slezine, operacija usmerena na očuvanje tkiva treba da bude glavni cilj. Otvorena parcijalna splenektomija kojoj su bili podvrgnuti i jedan i drugi bolesnik bila je bez intraoperativnih ili postoperativnih komplikacija i neometanog praćenja, te bi mogla biti jedna od sigurnih opcija lečenja.

Ključne reči: epitelne ciste, slezina, deca