# The management of primary nonparasitic splenic cysts

Özlem Boybeyi, İbrahim Karnak, F. Cahit Tanyel, Arbay Ö. Çiftçi, M. Emin Şenocak
*Department of Pediatric Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey*

**SUMMARY: Boybeyi Ö, Karnak İ, Tanyel FC, Çiftçi AÖ, Şenocak ME. The management of primary nonparasitic splenic cysts. Turk J Pediatr 2010; 52: 500-504.**

**Primary nonparasitic splenic cysts (PNSC) are rare and their management in children has been controversial. We conducted this study to discuss various treatment modalities. The medical records of patients with PNSC (1991-2008) were evaluated retrospectively, including age, sex, history of trauma, presenting symptoms, physical examination and radiological findings, therapeutic approaches, and outcomes. Six patients, between 3 to 12 years of age with a male/female ratio of 2, were included. The presenting symptom was abdominal pain in all but one asymptomatic patient. Physical examination findings were unremarkable in all except for palpable spleen in two patients. Cyst sizes ranged from 3 x 4 cm to 10 x 12 cm. The patients were treated with aspiration-sclerotherapy (n=2), total splenectomy (n=2), partial splenectomy (n=1), and cyst excision (n=1). The pathological diagnoses were epidermoid cyst (n=5) and lymphangioma (n=1). The postoperative course was uneventful except for postsplenectomy fever (n=1), recurrence (n=1) and residual cyst (n=1). PNSC larger than 5 cm in diameter or those that are symptomatic should be treated surgically. Total splenectomy should not be done in children to avoid infectious postsplenectomy problems unless there is a mandatory condition like intraoperative bleeding. Aspiration-sclerosis is not recommended because of recurrence.**

***Key words: splenic cyst, epidermoid cyst, lymphangioma, primary, nonparasitic.***

Primary nonparasitic splenic cysts (PNSC) are rare and account for 10% of all nonparasitic splenic cysts, but they are the most frequent type of splenic cysts in children. The number of diagnosed splenic cysts seems to have risen because of increased use of abdominal imaging by ultrasound (US)[1-3]. Several treatment modalities have been suggested in the English language medical literature, such as splenectomy, cystectomy, aspiration with sclerosis, and marsupialization. However, the optimal mode of management of PNSC is still obscure. Furthermore, there is no clear explanation about which method should be preferred in a certain condition.

Therefore, we conducted a retrospective study evaluating PNSC cases treated in our department to suggest the criteria for selection of the most appropriate mode of treatment in the management of PNSC in children.

## Material and Methods

The medical records of patients treated for PNSC between 1991 and 2008 were evaluated retrospectively. The clinical, radiological and surgical findings were reviewed. The age, sex, history of trauma, presenting symptoms and physical examination findings, radiological findings, type of management, and outcome were noted.

## Results

Six patients were enrolled into the study. Data are summarized in Table I and Table II. There were five cases of epidermoid cysts and one case of lymphangioma. Patients ranged in age from 3 to 12 years. The male to female ratio was 2. None of the patients had a history of trauma and/or infectious disease.

The presenting symptom was abdominal pain in five cases, while one case was asymptomatic. Laboratory data and physical examination

**Table I.** Summary of the Clinical and Radiological Features of Cases with Primary Nonparasitic Splenic Cysts

| Case no | Age (year) | Sex | Presenting symptom | Imaging method | Size (cm) |
|---|---|---|---|---|---|
| 1 | 3 | F | Asymptomatic | US | 5 x 4 |
| 2 | 8 | M | Abdominal pain | US, CT | 10 x 11 |
| 3 | 11 | M | Abdominal pain | US, CT | 7 x 8 |
| 4 | 8 | F | Abdominal pain | US | 10 x 12 |
| 5 | 10 | M | Abdominal pain | US, CT | 3 x 4 |
| 6 | 12 | M | Abdominal pain | US, CT | 10 x 12 |

F: Female. M: Male. US: Ultrasound. CT: Computerized tomography.

findings were unremarkable in all cases except for palpable spleen in two cases. All cases were also negative for echinococcus antibody.

The diagnostic radiological methods were abdominal US and/or abdominal computerized tomography (CT) in 6 and 4 patients, respectively. US revealed a round homogeneous anechoic area with thin septations and smooth thin wall. Contrast-enhanced CT scans demonstrated a spherical, well-defined lesion with attenuation near water and no rim enhancement without calcification (Fig. 1, 2). The cyst was solitary in all cases, with sizes ranging from 3 x 4 cm to 10 x 12 cm in diameter.

The initial diagnosis based on clinical features and imaging findings was PNSC. Two cases underwent total splenectomy (Cases 2 and 4). Case 2 had an attack of fever 10 days after splenectomy. He was discharged uneventfully following antibiotherapy. Case 5 had a partial splenectomy and his clinical course was uneventful. He has been under follow-up with US for four years without complaint. Percutaneous aspiration of the cyst and alcohol injection were applied in Case 1 and Case 3. The cyst of Case 1 disappeared within one month; however, she had an asymptomatic recurrent cyst with a diameter of 2 x 2 cm in three months. She has been under follow-up with US for 18 months since she has no complaints. Case 3 had an asymptomatic residual splenic cyst with a diameter of 2 x 1.5 cm. He has been under follow-up with US for 10 months also without complaint. Total excision of the cyst was performed in Case 6, and his clinical course was uneventful. He has had no cyst for four years.

Microscopic evaluation revealed that the wall of the epidermoid cysts appeared like a fibrous capsule with trabeculated internal surface. The cysts were lined by stratified squamous epithelium. The lymphangioma was a multilocular cyst with thin septations and its inner surface was lined by endothelium.

The duration of follow-up ranged from 1 to 48 months (mean: 21 months). Recurrent cyst was encountered in only one patient (Case 1), who had been treated with aspiration and sclerosis. Residual cyst was encountered in only one

**Table II.** Summary of Surgical, Pathological and Follow-Up Data of Cases with Primary Nonparasitic Splenic Cysts

| Case no | Management | Pathology | Outcome |
|---|---|---|---|
| 1 | Percutaneous drainage with sclerosis | Epidermoid cyst | Recurrence |
| 2 | Total splenectomy | Epidermoid cyst | No evidence of disease* |
| 3 | Percutaneous drainage with sclerosis | Epidermoid cyst | Residual cyst |
| 4 | Total splenectomy | Epidermoid cyst | No evidence of disease |
| 5 | Partial splenectomy | Lymphangioma | No evidence of disease |
| 6 | Total excision of the cyst | Epidermoid cyst | No evidence of disease |

*The patient had an attack of fever 10 days after splenectomy.

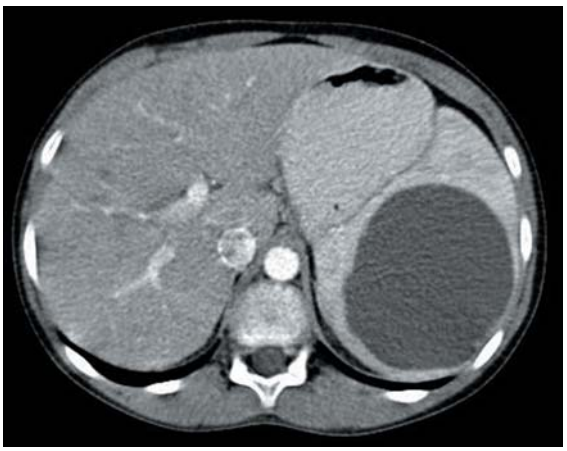

**Fig. 1.** CT scan of Case 3 revealing epidermoid cyst (7 x 8 cm) located centrally.

patient (Case 3), who had been treated with aspiration and sclerosis. The clinical course was uneventful in the remaining patients.

## Discussion

Splenic cysts are classified traditionally as primary or secondary cysts according to the presence or absence of an epithelial lining of the lumen[1-3]. Primary splenic cysts are further subdivided into parasitic or nonparasitic subgroups[1,3]. Secondary cysts are also referred to as posttraumatic or pseudocysts of the spleen. However, epithelial lining of primary cysts may be atrophic or desquamated, which sometimes makes it difficult to distinguish the primary cysts from secondary cysts[3-7]. In other words, absence of epithelial lining neither confirms the diagnosis of secondary cyst nor excludes the possibility of primary cyst. Nonparasitic cysts are also subdivided into two groups as congenital or neoplastic cysts. Congenital cysts are epidermoid and dermoid cysts. Lymphangioma and hemangioma are considered in the neoplastic group[1,3,7]. In the present study, five cases were epidermoid cysts and one was lymphangioma.

The incidence of splenic cysts is low. The PNSC are seen predominantly in children and young adults[1, 3,5,7]. The ages of our cases ranged from 3 to 12 years.

The PNSC are usually large in size. They are usually solitary but may be multiple. There are a few familial cases reported in the English medical literature[1,8,9]. In the present study, cyst size ranged from 3 x 4 cm to 10 x 12 cm in diameter, all cases were nonfamilial, and all cysts were solitary.

The clinical presentation differs according to the size of the cysts. It is more likely to be symptomatic if the cyst is large, as occurred in our series. The symptoms caused by the cyst are nonspecific, such as epigastric pain or fullness, and are best related to compression of the adjacent organs[1,3-7,10]. Rarely, reversible hypertension due to renal artery compression may be seen. Varicocele was also reported[11]. Complication of splenic cysts is seen rarely. However, rupture of cyst causing hemoperitoneum, peritonitis, abscess, anaphylactic shock, and empyema may be seen. Hypersplenism and secondary cyst infection have also been reported[1,3-5,10].

Although the laboratory data and physical examination findings are usually unremarkable, anemia, granulocytopenia and thrombocytopenia may be seen rarely. Splenomegaly or palpable mass may be encountered in the left upper quadrant on physical examination[1,3,10]. Serum carbohydrate antigen 19-9 (CA 19-9) and carcinoembryonic antigen (CEA) have been reported to be elevated in both serum and cyst fluid in patients with epidermoid cysts[12]. These antigens were not investigated in the present study, and the remaining laboratory data and physical findings were unremarkable except for palpable spleen in two patients.

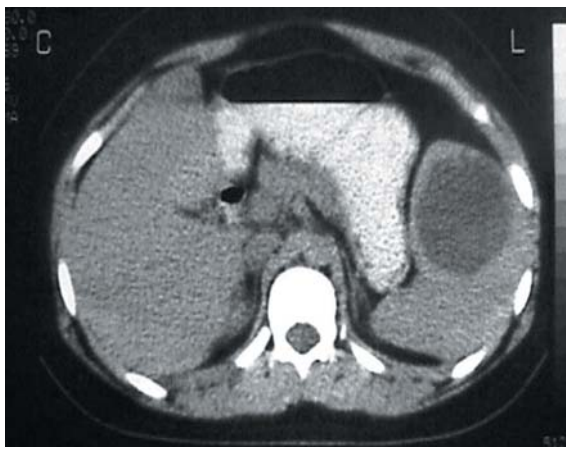


**Fig. 2.** CT scan of Case 5 revealing cystic lymphangioma (3 x 4 cm) located at the upper pole of the spleen.

Ultrasonography and CT are the mostly frequently used diagnostic methods. They are helpful in determining whether the cyst is multilocular or unilocular, the location of the cyst in the spleen and its relationship to the surrounding structures[1,3,10]. US is a sensitive, cheap and noninvasive tool in the diagnosis. PNSC appear as a round homogeneous, anechoic area with marked echo enhancement and with a smooth, thin wall in the US study. A more complex picture with accompanying irregular border, septation and calcification may be seen in some cases[1,3,5,6,11]. PNSC are spherical, well-defined lesions without rim enhancement on CT. Calcification of the cyst wall and septations are well demonstrated with CT[1,3,5,6]. US and CT were used in combination for the diagnosis in our series. All cysts were unilocular, having a well-defined border, with no calcification on the cyst wall.

The conventional treatment of PNSC for years was total splenectomy. It was determined later that the spleen is involved in several functions including regulation of circulatory blood volume, hematopoiesis and immunity, and that total splenectomy may expose the child to a risk of serious infectious complications such as postsplenectomy sepsis. The risk of postsplenectomy sepsis has been reported to be 4%, with a mortality rate of 1.5%[13]. Therefore, splenic preservation to the extent possible is recommended in children, and total splenectomy must be preserved as a last option unless there is excessive bleeding during partial splenectomy, total or near total replacement of splenic parenchyma by a large PNSC or the presence of an accompanying complication such as hypersplenism. The patient should be prepared for total splenectomy with vaccination against the encapsulated bacteria and antibiotic coverage during pre- and postoperative periods as well as long-term antibiotic prophylaxis. If the total splenectomy was not foreseen preoperatively, coverage with large- spectrum antibiotics and close monitoring are necessary in order to not overlook sepsis[1,3,10,11,13].

A number of treatment options, such as aspiration with or without sclerosis, internal or external marsupialization, partial splenectomy, cystectomy, and decapsulation were suggested later to avoid serious infectious complications following total splenectomy[1,4,5,10,14].

Additionally, nonoperative treatment has been recommended for small cysts up to 5 cm in diameter, if the cyst is completely asymptomatic and the imaging findings are absolutely typical of PNSC[4,7]. Percutaneous drainage of the cyst, with or without application of a sclerosing agent, is followed by a high incidence of recurrence. Additionally, dense inflammatory response around the spleen may make subsequent operation difficult[4,11,15]. Marsupialization is creation of an opening on the cyst wall for internal or external drainage of the cyst cavity. This technique is also not recommended because of recurrence risk[1,3,4]. The term splenic decapsulation, which is also a kind of internal drainage, defines near total resection of the cyst, leaving some portion of the wall retained. The recurrence of the cyst may be evident following this type of treatment[2,4,14,16].

Removal of the cyst wall with lining epithelium is the only treatment with definitive assurance that no cyst remnant remains[1,3,4,10]. Therefore, total excision of the cyst provides an acceptable therapy result. It can be tried in a superficial cyst that is not close to the splenic hilum. Partial splenectomy can be applied if the PNSC is located deep in the parenchyma or if the cyst is large. It can be performed with low risk in children. However, the location of the cyst should be suitable to perform partial splenectomy without jeopardizing the vascular support of the remaining spleen. It is usually performed in the transverse manner; however, it can also be performed safely in a vertical manner[17]. It should be kept in mind that partial splenectomy may cause excessive intraoperative bleeding that necessitates total splenectomy[3-6,11,13,14,18,19]. The surgeon should carefully control the bleeding splenic surface and not be resistant to conversion to total splenectomy if the bleeding cannot be controlled effectively.

The preferable treatment modality for small cysts less than 5 cm in diameter is close follow-up. Cysts larger than 5 cm in diameter or symptomatic cysts should be treated surgically. In spite of the limited number of cases, we absolutely recommend partial splenectomy or total removal of the cyst in suitable cases instead of total splenectomy. Total splenectomy should be avoided in children because of undue loss of some important functions of the spleen

in a child and probability of serious infectious complications in the postoperative period. It must be reserved for mandatory conditions such as uncontrolled bleeding during operation or for cases with total replacement of splenic tissue by a very large cyst. Aspiration and sclerosis is not recommended because of cyst recurrence.

**REFERENCES**

1. Abbasoğlu O, Kılıc YA. Splenic cysts. In: Sayek I (ed). Temel Cerrahi. Ankara: Güneş Kitabevi; 2004: 1450-1451.
2. Mackenzie RK, Youngson GG, Mahomed AA. Laparoscopic decapsulation of congenital splenic cysts: a step forward in splenic preservation. J Pediatr Surg 2004; 39: 88-90.
3. Hansen MB, Moller AC. Splenic cysts. Surg Laparosc Endosc Percutan Tech 2004; 14: 316-322.
4. Morgenstern L. Nonparasitic splenic cysts: pathogenesis, classification and treatment. J Am Coll Surg 2002; 194: 306-314.
5. Czauderna P, Vajda P, Schaarschmidt K, et al. Nonparasitic splenic cysts in children: a multicentric study. Eur J Pediatr Surg 2006; 16: 415-419.
6. Dachman AH, Ros PR, Murari PJ, Olmsted WW, Lichtenstein JE. Nonparasitic splenic cysts: a report of 52 cases with radiologic-pathologic correlation. AJR 1986; 147: 537-542.
7. Musy PA, Roche B, Belli D, Bugmann P, Nussle D, Le Coultre C. Splenic cysts in pediatric patients -- a report on 8 cases and review of the literature. Eur J Pediatr Surg 1992; 2: 137-140.
8. Younger KA, Hall CM. Epidermoid cyst of the spleen: a case report and review of the literature. Br J Radiol 1990; 63: 652-653.
9. Ragozzino MW, Singletary H, Patrick R. Familial splenic epidermoid cyst. AJR 1990; 155: 1233-1234.
10. Rescorla FJ. Spleen. In: Grosfeld JL, O'Neill JA Jr, Coran AG, Fonkalsrud EW (eds). Pediatric Surgery. Philadelphia: Mosby-Elsevier; 2006: 1692.
11. Tsakayannis DE, Mitchell K, Kozakewich PW, Shamberger RC. Splenic preservation in the management of splenic epidermoid cysts in children. J Pediatr Surg 1995; 30: 1468-1470.
12. Arda IS, Tüzün M, Hiçsönmez A. Epidermoid cyst of the spleen with elevated levels of CA125 and carcinoembryonic antigen. Eur J Pediatr 2005; 164: 108.
13. Kimber C, Spitz L, Drake D, et al. Elective partial splenectomy in childhood. J Pediatr Surg 1998; 33: 826-829.
14. Mertens J, Penninckx F, De Wever I, Topal B. Long-term outcome after surgical treatment of nonparasitic splenic cysts. Surg Endosc 2007; 21: 206-208.
15. Carpenter G, Cotter PW, Davidson JR. Epidermoid cyst of the spleen. Aust NZ J Surg 1986; 56: 365-368.
16. Schier F, Waag KL, Ure B. Laparoscopic unroofing of splenic cysts results in a high rate of recurrences. J Pediatr Surg 2007; 42: 1860-1863.
17. Golinsky D, Freud E, Steinberg R, Zer M. Vertical partial splenectomy for epidermoid cyst. J Pediatr Surg 1995; 30: 1704-1705.
18. Kimber C, Pierro A, Drake D, Kiely E, Spitz L. Hemisplenectomy for giant splenic cysts in children. Pediatr Surg Int 1998; 14: 116-118.
19. Pappis CH, Demitriadis D, Petrou A, Pappis HC. Postoperative recurrence of splenic epithelial cyst. Treatment with subtotal splenectomy. Z Kinderchir 1990; 45: 245-248.