Other full case

Mesothelial cyst of the spleen in an adult: a case report

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Summary

A young adult in her third decade presented with a 2-week history of catching left upper abdominal pain and was detected to have a cystic lesion occupying almost the entire spleen. Laparoscopic total splenectomy was carried out, and the cyst wall revealed a true mesothelial cyst with no squamous metaplasia. The various aspects of mesothelial cysts, including immunophenotyping and treatment modalities, are briefly discussed.

BACKGROUND

Non-parasitic cysts of the spleen are rare and are incidentally discovered either on imaging or while investigating for a left upper abdominal pain. Derived from the mesothelial cell lining of the splenic capsule,¹ these uncommon true cysts may remain asymptomatic for a long time² and have a

characteristic gross and microscopic appearance.^{1 2} The lining cells stain positive for calretinin, a mesothelial marker, and secrete CA 19-9 which can be used as a screening tool in the management of these lesions.^{3 4} Our case is a typical example of these rare, true, non-parasitic cysts of the spleen.

Figure 1 Splenic cyst with smooth, glistening inner wall with typical tree-root trabeculations.

Figure 2 Splenic cyst wall lined by cuboidal mesothelial cells (H&E, ×400).

CASE PRESENTATION

A young lady in her third decade presented with acute onset of catching left flank and upper abdominal pain of 2 weeks duration. She denied any other significant complaints. Clinical examination was unremarkable except for a moderately enlarged, slightly tender spleen.

INVESTIGATIONS

Laboratory investigations were essentially normal. An abdominal ultrasound examination showed features of a splenic cyst; CT of the abdomen confirmed the presence of a splenic cyst with a differential diagnosis of false cyst, lymphangioma and hydatid disease being considered. Laparoscopic findings included a large cyst occupying the splenic hilum precluding dissection of the vascular pedicle for partial splenectomy or marsupialisation. An uneventful laparoscopic splenectomy was performed.

Grossly, the spleen measured 14×11×6 cm, with the splenic cyst measuring 12×8×4 cm. The cyst was filled with brown coloured fluid. Cut section showed a smooth, glistening inner wall with typical tree-root trabeculations (figure 1). The cyst wall was lined by flattened cuboidal mesothelial cells (figure 2). Immunophenotyping showed positivity for calretinin (figure 3) throughout the lining epithelium, positivity for low molecular weight cytokeratin and a weak positivity for CA 19-9 in the wall. Based on

the histological and immunophenotypic findings, a final diagnosis of primary mesothelial cyst of the spleen was made.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis include a false cyst, parasitic cyst or occasionally, a lymphangioma.

TREATMENT

An uneventful laparoscopic splenectomy was carried out; initial attempts to preserve the spleen were abandoned due to the central location and large sized cyst, precluding vascular dissection.

OUTCOME AND FOLLOW-UP

One year postsurgery, she is asymptomatic and has no complaints.

DISCUSSION

Ever since the first reported case of splenic cyst by Andral in 1929, the classification of these lesions has evolved into the present system.⁵ Broadly classified as parasitic and non-parasitic cysts, the non-parasitic are further categorised as primary (epithelial/true) and secondary (false/pseudo) cysts based on the lining of the cyst.⁶ Parasitic cysts are generally

Figure 3 Mesothelial lining showing strong positivity for calretinin (IHC, $\times 400$).

seen in endemic areas and are usually caused by *Echinococcus granulosus* infestations.

The true or primary cysts may be congenital or neoplastic in origin and are lined by mesothelial, squamous or transitional epithelium. Secondary or pseudocysts are usually post-traumatic, due to failure of organisation of subcapsular or parenchymal haematomas and occasionally due to necrosis following an infarction or rarely due to an abscess.⁵

Clinically, primary cysts occur predominantly in children and young adolescents; often asymptomatic until they assume large sizes, they may then present with local or referred pain, abdominal distension, compression of adjacent structures and rarely as thrombocytopenia.^{1 6 7}

Capsular surface mesothelial invagination with subsequent cyst formation,⁶ embryonic inclusion of epithelial cells from adjacent structures, epithelial cell metaplasia from adjacent structures or vascular endothelium from peritoneal inclusions are some of the theories put forth to explain the genesis of these congenital cysts.¹

Grossly, mesothelial are usually unilocular and vary in size; the cut surface is whitish or greyish-white, smooth and glistening and displays characteristically prominent coarse tree-root-like trabeculations due to subepithelial fibrosis. Cyst fluid may vary in density and may demonstrate cholesterol crystals and macrophages on microscopic examination.⁸

The cyst wall is generally lined by a single cell series of mesothelial, transitional or squamous epithelium.^{1 8} Occasional finding of combination of epithelial cells may be explained by the pluripotent nature of the mesothelial cells.² Occasional absence of lining cells may lead to an erroneous diagnosis of secondary cyst unless multiple sections are studied.⁵

Accurate preoperative diagnosis of mesothelial cysts is rare; the occurrence of a unilocular cyst in the absence of previous trauma, infection or exposure to hydatid disease may help in the diagnosis.⁹

Recent studies have indicated rise in tumour markers such as CA 19-9 and carcinoembryonic antigen levels in association with primary mesothelial cysts.³ Serum levels of CA 19-9 have been shown to reduce substantially after cyst removal, offering a screening test to indicate recurrences in case of spleen preserving surgeries.³ CA 19-9 may also be demonstrated in lining of the cyst wall as was demonstrated in our case.⁴ Calretinin is a vitamin D-dependent calcium-binding protein that is expressed in mesothelial cell lines; even benign mesothelial cells may show the characteristic “fried-egg” staining pattern.¹⁰ Cytokeratins specifically permit identification of the epithelial origin based on the expression profile. Immunohistochemistry confirmed the mesothelial origin of the splenic cyst in our case as evidenced by the positive calretinin and low molecular weight

cytokeratin and weakly positive CA 19-9 staining in the cyst wall.

All symptomatic cysts and cysts beyond 5 cm in diameter need surgical treatment. Splenic preservation should be the primary factor while considering treatment; in the present era, laparoscopic methods are increasingly being carried out. Various modes of treatment based on the expertise of the surgical team include total splenectomy, partial cyst excision and marsupialisation,⁷ partial splenectomy, partial cystectomy (decapsulation) and laparoscopic partial cystectomy.⁸ Our patient was subjected to a laparoscopic splenectomy because the cyst was occupying the hilar portion precluding preservation.

In conclusion, a rare case of histologically and immunophenotypically confirmed mesothelial cyst of the spleen is presented with a discussion of the various findings and treatment modalities.

Learning points

- ▶ True or primary splenic cysts are rare and are generally incidentally discovered on an ultrasound examination or CT of the abdomen.
- ▶ Mostly asymptomatic, some of them present with left upper abdominal or flank pain.
- ▶ Laboratory investigations are essentially normal.
- ▶ Surgical management of symptomatic cysts and cysts beyond 5 cm in diameter should be considered as a first option with splenic preservation being the prime consideration.
- ▶ Surgery offers the best curative and least recurrence rates. Marsupialisation, fenestration, partial deroofing, partial cystectomy or splenectomy can all be considered and carried out laparoscopically. Occasionally, total splenectomy may be required.

Competing interests None.

Patient consent Obtained.

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