

Case Report

A primary splenic epithelial cyst: rare case report

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ABSTRACT

Splenic cysts are rare, most of them being hydatid cysts. Our case is about a primary (true) splenic epithelial cyst. A 21-year-old female patient presented to casualty with pain in the upper left abdomen. On being investigated with sonography she was found to have a solitary cystic lesion in the spleen. Her contrast enhanced computed tomography of the abdomen and pelvis revealed a 10.9 × 11.4 × 12.0 cm cystic lesion of the spleen, probably a hydatid or primary splenic epithelial cyst. Due to the large size of the cyst, an open surgical approach was planned. On exploratory laparotomy via a left subcostal incision, a solitary cyst involving >75% of the splenic parenchyma was discovered. Total splenectomy was done as splenic parenchyma could not be functionally salvaged. Histopathological investigation showed a primary epithelial splenic cyst. The patient recovered uneventfully.

Keywords: Cyst, Epithelial, Primary, Spleen, Splenectomy

INTRODUCTION

Primary splenic cysts are unusual and often an incidental finding in surgical practice. As per the existing literature, since the first case was reported in 1929 by Andral, the classification of these lesions has evolved into the present system. Splenic cysts may be parasitic or non-parasitic in origin. Nonparasitic cysts are either primary or secondary. Primary cysts are also called true, congenital, epidermoid or epithelial cysts. Primary splenic cysts account for 10% of all benign non-parasitic splenic cysts are congenital and the most frequent type of splenic cysts in children.¹ Hydatid cysts are the commonest of these cysts. Generally, splenic cyst is asymptomatic and do not grow to more than 4 cm in size. The symptoms are related to the size of cysts. They may present with fullness in the left abdomen, local or referred pain, symptoms due to compression of adjacent structures (like nausea, vomiting, flatulence, diarrhoea) or rarely thrombocytopenia, and occasionally complications such as infection, rupture and/or haemorrhage. Contrast

enhanced CT of the abdomen is the best investigation for these cysts. However, careful histopathological evaluation along with immunostaining for presence of epithelial lining is mandatory to arrive at the diagnosis. The treatment has changed drastically from total splenectomy in the past to splenic preservation methods recently. Smaller cysts are managed conserving the splenic parenchyma, while larger cysts (>4-5 cm) may require splenectomy.²

CASE REPORT

A 21-year-old female patient presented with pain in the left upper abdomen for 15 days which was dull aching and increased in intensity after meals. There was no history of trauma or contact with any animals. On examination abdomen was soft and non-tender. The spleen was just palpable. Abdominal X-rays showed an absence of a normal fundic shadow. Ultrasonography of the abdomen revealed a giant cystic lesion in the spleen with calcifications; most probably a splenic hydatid.

CECT abdomen confirmed a large well defined non-enhancing cystic lesion involving the superior mid and lower part of the spleen of size 10.9 × 11.4 × 12.0 cm. The spleen was abutting the fundus of the stomach medially and was displacing the left kidney inferiorly. (Figure 1).



Figure 1: (a) sections from the abdomen showing large cystic lesion in the splenic parenchyma occupying almost whole of the spleen. (b) coronal sections of CECT abdomen and pelvis showing left kidney displaced downwards due to huge splenic cyst.

Open laparotomy was planned due to large size of the spleen. At surgery, a large 20 × 15 × 10 cm cyst was found occupying more than 75% of the spleen, hence total splenectomy was done (Figure 2).



Figure 2: (a) intraoperative snapshot of large splenic cyst occupying almost all of the splenic parenchyma. (b) needle aspiration of cystic fluid showing brownish fluid aspirate.

Histopathology report confirmed a large uni-locular cyst 20 × 14 × 10 cm with brown coloured serous fluid. Microscopy revealed cyst wall lined by low cuboidal epithelium with focal stratification, which is a rare presentation. There was no evidence of malignancy.

Thus, the diagnosis of primary true splenic epithelial cyst was established.

DISCUSSION

Splenic cysts are classified based on the presence or absence of an epithelial lining, etiology, pathogenesis, by Fowler and Martin based on the cellular lining of the cystic wall, as primary (true) and secondary (pseudo) cysts.^{3,4} They may also be classified as parasitic or non-parasitic. A new classification based on the true pathogenesis of cyst divides non-parasitic splenic cysts as congenital, neoplastic, traumatic, and degenerative.⁵

Congenital splenic cysts are called epidermoid or epithelial cysts. They are uncommon, comprising only 10% of all splenic cyst and 25% of non-parasitic cysts. The pathogenesis of primary splenic cysts is not clear. In view of this, many hypotheses were proposed. Mesothelial invagination theory postulated that in case of congenital cysts, it is postulated that during development there is invasion of mesothelial lining along with the capsule. As the lining has pluripotent nature, it has propensity to undergo metaplasia and secretion of fluid, leading to the formation of cysts.⁶ Another postulation is that they form from normal lymph spaces within the spleen.^{6,7} Endodermal inclusion theory proposes that epithelial splenic cysts develop by true metaplasia of a heterotopic endodermal inclusion within the spleen.⁸

Splenic epithelial cysts occur predominantly in the 2nd and 3rd decades of life but can occur in children and infants. They are more common in females.^{8,9} Small cysts are usually asymptomatic. The most common presentation is an asymptomatic lump in the abdomen. Mass effect can also give rise to vague symptoms like abdominal discomfort, early satiety, nausea and dysphagia.¹⁰ Diagnosis is best made by CT scan of the abdomen. It gives information regarding the morphology of the cyst, composition of cystic fluid, location of the cyst within the spleen and its relationship with surrounding tissues. The serum tumour markers carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA 19-9) may be raised and so checked.¹¹ However, definite diagnosis is possible only after splenectomy when epithelial lining is confirmed by histopathology along with immunohistochemistry. Histologically, epidermoid cysts of the spleen have a squamous or low cuboidal lining with intracellular bridges and a thick collagenous wall. The cystic fluid may contain cholesterol crystals, protein particles or breakdown products of haemorrhage. The mesothelium undergoes metaplasia to squamous epithelium secondary to chronic irritation.¹²

In earlier days open complete splenectomy was the preferred treatment of most of the splenic cysts. Over the years the trend has shifted to more conservative management due to the demonstration of increased mortality of splenectomised patients due to

overwhelming post-splenectomy sepsis.¹³ Cysts occupying more than 2/3rd of the splenic volume are best treated by complete or partial splenectomy.¹⁴ Laparoscopic surgery or laparotomy can be done as per the surgeons and patient factors. The important thing is to conserve at least 25% of the splenic parenchyma. This is the minimal tissue required to preserve the immunological functions of spleen, without increasing the risk of recurrence.¹⁵

CONCLUSION

An accurate preoperative diagnosis of primary epithelial cysts is difficult; the occurrence of a cystic lesion in the absence of previous trauma, infection or exposure to hydatid disease may help to arrive at the diagnosis. However, careful histopathological evaluation along with immunostaining is important to arrive at correct diagnosis. The treatment has changed drastically from total splenectomy in the past to splenic preservation methods recently to preserve the splenic function in order to avoid untoward complications related to historical treatment.

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