Case report:

Splenic Epidermoid Cyst - A Rare Case Managed in a private medical college hospital of Bangladesh

Md Jahangir Hossain Bhuiyan[1], Mohammad Mohibul Aziz[2], Md Omar Faruk[3], Md Mahbub Hasan[4]

Abstract

Splenic cysts are rarely detected, and clinically diagnosed commonly as spleenomegaly . The nature of the cyst could not be diagnosed pre-operatively because of its' anatomical location and only be confirmed by the histopathological assessments. These cysts are found as a sequel of splenic infarct followed by the abscess formation. This is a very rare condition of a minor boy of only 10 years' old who presented with a hugely enlarged spleen. The common investigation(s) did not prove that it was in the haematological conditions. Ultrasonogram (USG) investigation shows spleenomegaly due to the cystic lesion. CT scan of the abdomen showed large and well defined cystic mass which was 13.4cmX7.9cmX15.3cm in size, multi-loculated, septated, sharply demarcated as such well delineated cyst. After necessary preparation like vaccination, blood grouping, cross matching and other fitness, laparotomy and total spleenectomy was done as the cyst occupied most of the Splenic tissue. The post operative recovery was uneventful and the final diagnosis was confirmed by histopatholgical examinations as epidermoid cyst of the spleen. The patient was followed up for two consecutive months and not a single complication was noticed.

Keywords: Spleen; Cystic Lesions; Epidermoid cyst

Bangladesh Journal of Medical Science Vol. 20 No. 01 January'21. Page : 177-180
DOI: https://doi.org/10.3329/bjms.v20i1.50366

Introduction

The non-parasitic splenic cysts are very rare case. Splenic cysts are classified as primary cysts (true) or pseudocysts (secondary) on the basis of presence or absence of lining epithelium. True cysts form embryonal rests and include dermoid and mesenchymal inclusion cysts. True cysts of the spleen are very uncommon, rare and usually classified as the cystic hemangiomas, cystic lymphangiomas and epidermoid and dermoid cysts.

Epidermoid splenic cysts are the example of primary congenital cysts that contain an epithelial lining, unlike secondary cysts, which are composed of the fibrous tissues[1,2]. They are thought to be of the congenital origin and represent 10 per cent of the splenic cysts. They may be neoplastic, vascular, inflammatory and posttraumatic in origin and may contribute up to 30-40% of the total splenic lesions[3]. The congenital splenic cysts are commonly asymptomatic and they are rarely noticed in the routine surgical practice. It is due to lack of the typical as well as clinical presentation also, and they may be discovered very incidentally.

These should be differentiated from false or secondary cysts that they may result from trauma and contain serous or haemorrhagic fluid. The walls of such degenerative cysts may be calcified and therefore, resembled to radiological appearances of a hydrated cyst[4]. The splenectomy or partial splenectomy is usually considered for the cysts larger than 5 cm in diameter[4]. Presently, the emphasis is being laid on conservation of spleen, especially in the children and young adults, in order to avoid overwhelming post-

1. Md. Jahangir Hossain Bhuiyan, Professor
2. Md. Mohibul Aziz, Professor & Head of the Department, and Principal
3. Md. Omar Faruk, Assistant Professor
4 Md. Mahbub Hasan, Registrar
Department of Surgery, Ibn Sina Medical College, Kallyanpur, Dhaka, Bangladesh

Correspondence to: Professor (Dr) Md Jahangir Hossain Bhuiyan, Professor, Department of Surgery, Ibn Sina Medical College, Kallyanpur, Dhaka, Bangladesh.

splenectomy infections[5]. However, not all splenic cysts can be treated by partial splenectomy [6].

We are reporting a case of the large splenic epidermoid cyst - a very rare and very uncommon type of splenic cyst, which was removed by open spleenectomy totally highlighting the clinical presentation and the management option for such a kind of the patient in an economic poor resource setting in a larger portion of the population in Bangladesh.

Case Report

A 10-year-old male child presented with the chief complaints of mild dull dragging pain, discomfort and feeling of fullness in the left upper abdomen, which had been presenting for the past three months. There was no history of fever, trauma and associated with food intake. On physical examination, a firm, smooth, slightly tendered mass was seen in the left hypochondrium, which was palpable. He is non-diabetic, normotensive and non-asthmatic. He has not any positive family history relevant to this disease.

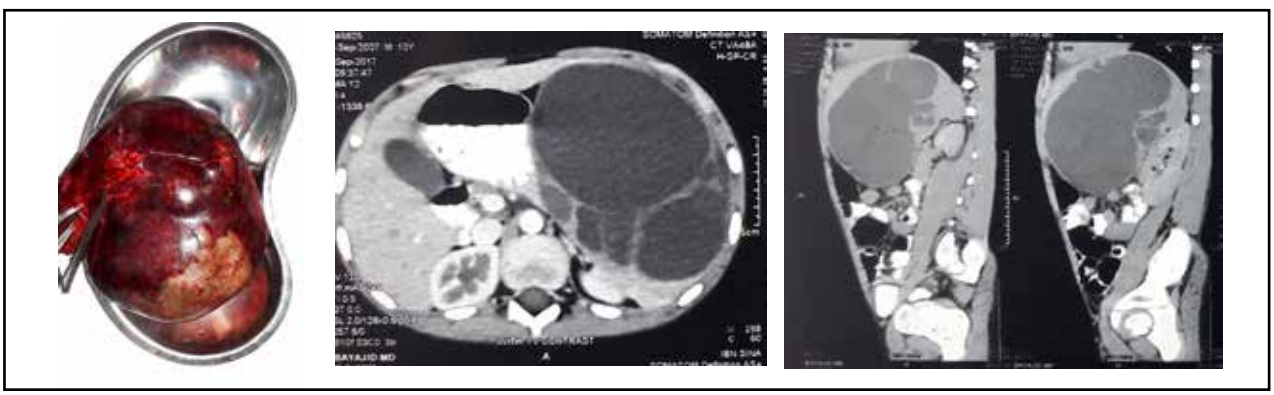
Laboratory investigations reveal that the Complete Blood Count showed normal findings. Hb elecreopheresis also reveals normal. HBsAg was negative. USG of the abdomen was done and it showed enlarged spleen with irregular cystic lesion, which measured 13cmx10cmx8cm in size, associated with an approximate 600 ml of volume also with thick mobile echogenic debris. X-ray of chest revealed slightly raised left hemidiaphragm. CT scan of the abdomen was conducted on a spiral scanner, after administering oral and intravenous contrast. Axial sections showed a large well defined cystic mass which was 13.4cmx7.9cmx15.3cm in size, multi-loculated, septated, sharply demarcated and well delineated cyst.

Elective laparotomy was performed and a large splenic cyst was found, which was located at the lower pole of the spleen. Due to location and size of the cyst, preservation of the spleen was considered impossible and splenectomy was done. Grossly, large sized cystic lesion which was approximately 10 cms of maximal diameter was discovered, with almost total replacement of splenic tissue at one side. Histopathological examinations determined that the whole cyst wall which was constituted by stratified squamous epithelium underneath the epithelium was fibrous tissue. Postoperatively he had an uneventful recovery from anesthesia. There was no other complication of wound. Stitches were removed on the 10th POD and the wound was healthy.

Histopathology shows unremarkable splenic parenchyma and cyst walls lined by the stratified squamous epithelium. Pathological diagnosis indicated a splenic epidermoid cyst.

Discussion

The splenic cystic lesions are very rare, uncommonly encountered in the routine surgical practice. Robbins reported a series of 42,327 autopsies over a 25-year period and results showed that only 32 patients had the diagnosis of the splenic cyst[4]. The classification, diagnostic modalities and treatment guidelines for splenic cysts are far from the uniform or clear. Only three classifications have been found in literature (Fowler's, Martin's and Morgenstern's classifications). Martin et al., had classified splenic cysts as Type-I cysts which were primary (true) cysts with cellular linings of the parasitic or non parasitic origin. Non-parasitic type-1 splenic cysts have been further divided into the congenital and neoplastic cysts. Congenital cysts include epidermoid, dermoid and endodermoid cysts, whereas the neoplastic cysts constitute hemangiomas, lymphangiomas. Among these, hemangiomas are the most common cases and dermoid cysts are the least commons. Type-II cysts are false (secondary) cysts without the cellular linings[5]. The differential diagnoses include intra-splenic abscesses, hydatid cysts, the true cystic neoplasms (hemangiomas, lymphangiomas,



epidermoid and dermoid cysts) and cystic metastases. Epidermoid cysts are the rarest among all true cysts which occur in the spleen[2]. Primary (true) cysts make up approximately 20% of all types of the cysts which occur in the spleen. The epidermoid cysts are the rarest, which comprise only 10% benign, non-parasitic cysts which occur in the spleen. Splenic epidermoid cysts are "true" cysts, as they possess inner epithelial linings of stratified squamous epithelium, whereas "false" cysts have no cellular linings, and are usually related to prior trauma, haemorrhage or infarction[4]. The origin of true cysts is still not very clear, but they are thought to develop from the mesonephric tissue in the developing spleen in early embryonic life. The mechanism which has been suggested is in-folding or entrapment of peritoneal mesothelial cells in the splenic parenchyma during embryogenesis in the intrauterine life. The age at presentation ranges from newborns to 50 years, with an average age in and around 17 years. There is slight female preponderance[6] Majority of the cases (near about 80%) present with solitary and unilocular cysts. The true cysts and false cysts are usually indistinguishable on imaging studies; false cysts tend to have thicker fibrous walls and eggshell like calcifications, along with internal debris. On USG, the epidermoid cysts are recognized as well-defined, thin walled, anechoic cystic lesions. There can be increased ecogenecity caused by the intracystic haemorrhage and cholesterol crystals and inflammatory necrotic debris which are same as those which are seen in false cysts. Although the peripheral calcifications occur more frequently in post-traumatic or false cysts, in epidermoid cysts with occasional internal septations, cystic walls may show curvilinear or plaque like calcifications. In addition, computerized tomography may give clearer picture regarding intracystic fluid, internal septations or calcifications. Besides, the partial splenectomy offers a definite solution in case of splenic cysts (<5 cm), conservative treatment like aspiration alone, incision and drainage can also be considered[7,8]. But total splenectomy is the treatment of choice in case of larger cysts (>5 cm), as it prevents the serious complications like rupture, haemorrhage, infection and sometimes, the recurrence which is attributed to coexistence of invisible tiny cysts that had remained in the splenic tissue after the dissection of the major cyst. In the cases where hilum is involved, the splenectomy remains a relatively safe procedure[3,7]. Finally, radiologically distinguishing the true and false splenic cysts does not seem to be possible preoperatively and the final diagnosis relies on the histopathological examinations of the wall of the cyst. However, the total splenectomy (open or laparoscopic) is primarily recommended for the prevention of complications which are seen as of the infections, haemorrhage and the rupture.

Conclusion

It is very important to recognize that the epidermoid splenic cysts may not be always present in a classic fashion on imaging so that it is trivial to include them as a differential diagnosis when warranted.

The Radiological examinations, particularly USG and CT scan can diagnose the splenic cysts unquestionably, correctly defining the relationships with adjacent organs. Splenomegaly or a splenic mass of a predominantly cystic nature with no clinical evidence of echinococcus suggests the diagnosis of the splenic cyst. The reliable radiological distinction between the true or false splenic cyst does not seem possible. CT scan and USG helps in detecting septa or the calcifications, which are definitely useful findings to distinguish the true from false cysts, since internal septa are more frequent in true cysts while parietal calcifications are typical of pseudocysts. The final diagnosis, however, is made at the histology. The exact nature of a cyst will not be known prior to histological examinations as in our case when imaging is not accurate or definitive in differentiation. Therefore, the cyst was treated based on symptomology, size, and location and a total splenectomy which was indicated.

Finally, surgery is primarily recommended in both the true and false large cysts for prevention of complications as of the infection, hemorrhage, and rupture thereof.

Ethical clearance: This case study was approved by local ethic committee.

Conflict of interest: None

Author's contribution:

Data gathering and idea owner of this study: Bhuiyan MJH, Aziz MM, Faruk MO, Hasan MM

Study design: Bhuiyan MJH, Aziz MM, Faruk MO, Hasan MM

Data gathering: Bhuiyan MJH, Aziz MM, Faruk MO, Hasan MM

Writing and submitting manuscript: Bhuiyan MJH

Editing and approval of final draft: Bhuiyan MJH, Aziz MM, Faruk MO, Hasan MM

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