Giant epithelial nonparasitic splenic cyst a pre-operative diagnosis dilemma: A case report

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SUMMARY

Giant splenic cyst is rare disorder affecting the spleen. As the occurrence is so in-frequent that the diagnosis preoperatively remains a challenge. We report a 12-year-old boy who presented to Sarawak General Hospital, Malaysia with left upper abdominal pain initially mistaken as a complex left liver cyst. He underwent surgery which turned out to be a giant splenic cyst and underwent laparotomy and total splenectomy. He was discharged well and remains asymptomatic after 6 months postoperative follow up.

INTRODUCTION

True splenic cyst is a rare clinical entity and is classified into parasitic and nonparasitic cyst.[1] Nonparasitic splenic cyst (NPSC) comprises of less than one third of all splenic cyst while true NPSC only makes up 10% of all benign splenic cysts.[1] Giant splenic cyst is defined as a cyst with a diameter greater than 15cm.[2] As parasitic splenic cyst is rare in Malaysia and NPSC occurrence is infrequent the diagnosis preoperatively remains a challenge. We report a case of a 12-year-old boy with a giant epithelial NPSC. To the best of our knowledge, this is the first giant epithelial NPSC to be reported in Malaysia.

CASE REPORT

A 12-year-old boy presented to a district hospital in Sarawak with left upper abdominal pain and distention for 1 month. Physical examination revealed a huge palpable left upper quadrant mass extending to the umbilicus. Contrast-enhanced computerized tomography scan (CECT) of the abdomen showed a large, well demarcated, cystic lesion at the left hypochondrium measuring 17cm x 15cm x 15cm inseparable from left lobe of liver and spleen with no associated calcification (Figure 1A). The rest of the cyst was homogeneously hypo-dense with attenuation suggestive of proteinaceous content (Hounsfield Unit 20) with a septum in between and irregular margins (Figure 1B). Serology for *Echinococcus granulosus*, Meliodosis, biohazard screening and tumour markers were all negative. The patient never had fever and other blood parameters were normal. Pre-operative diagnosis of an exophytic, complex liver cyst arising from segment II of liver was made. As our centre is a *Hepato-Pancreatico-Biliary center*, the child was referred to us and laparotomy and removal of cyst were discussed with the parents of the boy with possible left hemihepatectomy or splenectomy. The child was vaccinated prior to surgery with 3 intramuscular vaccines. The vaccines were the Pneumovax 23 a polyvalent pneumococcal vaccine (Merck Sharp & Dohme Corp, Sumneytown Pike, USA), Hiberix a polysaccharide conjugate *Haemophilus influenzae* type b vaccine (GlaxoSmithKline, Rixensart, Belgium) and Menactra a polysaccharide Diphtheria Toxoid conjugate vaccine (Groups A, C, Y and W-135) (Sanofi Pasteur, Swiftwater, USA). Intra-operatively, however the cyst was found to arise from the medial aspect of the spleen occupying almost the entire spleen while the liver was normal (Figure 2A). Total splenectomy was performed due to the large size of the cyst occupying the entire spleen (Figure 2B) The post-operative course was uneventful and he was discharged 5 days later. Pathological examination showed a unilocular epithelial splenic cyst with fibrous cyst wall (Figure 2C) lined by cuboidal cells (Figure 2D). At a follow-up review of 6 months, he was well.

DISCUSSION

Splenic cyst was first reported by Andral in 1929. The incidence was 0.07% in a large case series of 42327 autopsies over a 25-year period.[3] NPSC are mostly seen in children particularly females. NPSC is common in Europe and North America, while parasitic cyst is common in Africa and Central America.

The 2 main principal classifications by Fowler and Martin are based on the presence of cyst lining.[1,4] Type I cysts are primary (True) cyst with epithelial lining while Type II (False) cyst is without epithelial lining. True cysts can be further classified into parasitic and non-parasitic. In 2002, Morgenstern subdivides NPSC into congenital, neoplastic, traumatic and degenerative.[5] 80% of NPSC typically originates from trauma and degeneration while only 10% are congenital.

The pathogenesis is uncertain but there are 3 postulated theories. The first theory is the invasion of mesothelial lining after the rupture of the splenic capsule during development. This lining is pluripotent in nature and can undergo metaplasia with secretion of fluid leading to cyst formation.

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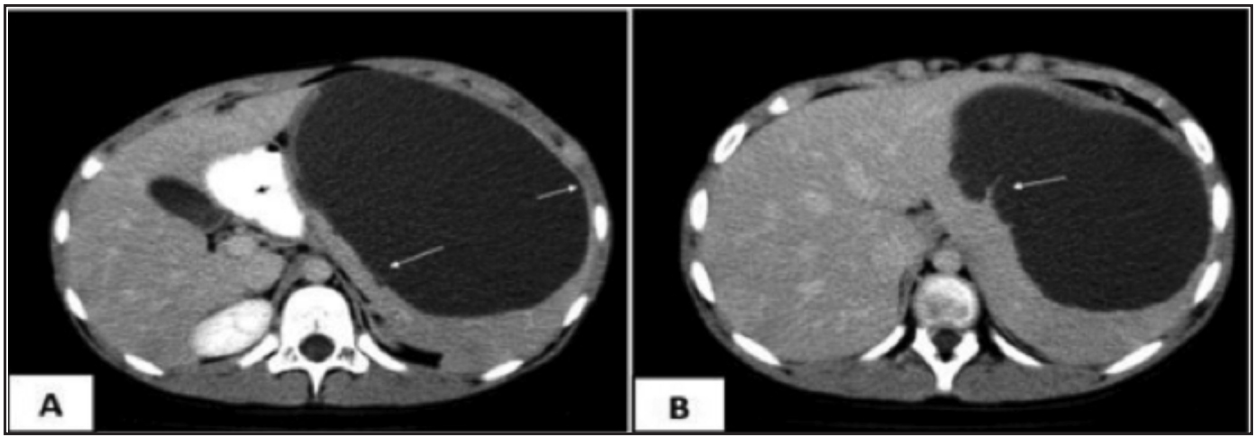

Fig. 1: (A) CECT showing large splenic cyst surrounded by splenic parenchyma posteriorly and anterolaterally (white arrow). Stomach is being pushed to the right (black arrowhead). (B) CECT scan showing thick septae (white arrow) and mild irregular wall at superomedial aspect of cyst.

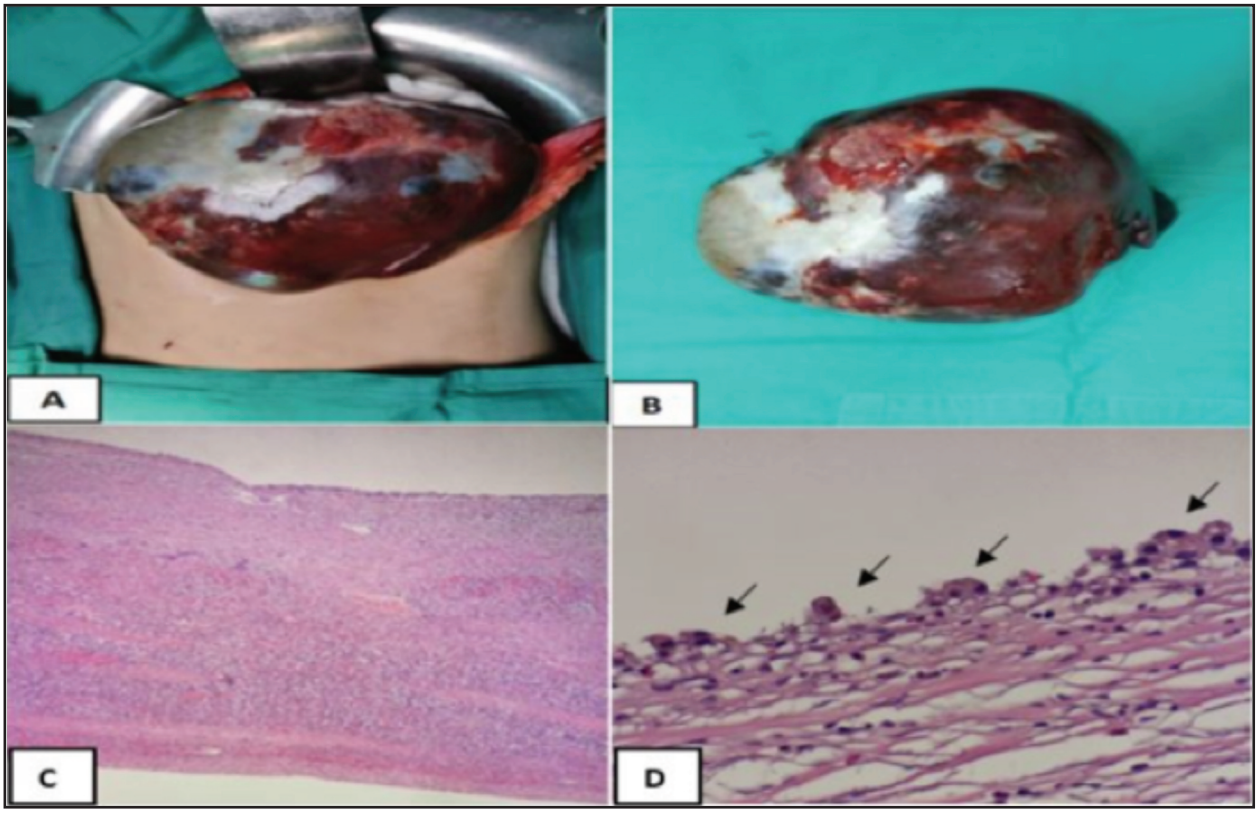

Fig. 2: (A) Giant splenic cyst intra operatively arising from the medial aspect of the spleen occupying the entire parenchyma. (B) Giant splenic cyst measuring 17cm x 15cm x 15cm removed intact together with the spleen. (C) Hematoxylin-Eosin stain: Fibrous wall infiltrated by lymphocytes (4x magnification). (D) Cyst wall lined by cuboidal cells (black arrows) (40x magnification).

The second theory is lymph space theory where the cyst arises from the normal lymph spaces in the spleen. The final theory is endodermal inclusion that proposes that epithelial splenic cyst develops by true metaplasia of the heterotopic endodermal inclusion within the spleen similar to the first theory. But there are studies that report that the epidermoid nature is due to the teratomatous differentiation or inclusion of fetal squamous lining instead of metaplasia.

Splenic cysts are usually diagnosed incidentally unless they are large enough to cause mass effect or complications. Various imaging modalities could be used to diagnose their cystic nature with ultrasound being the initial imaging modality of choice. For large lesion such as our case, cross-sectional imaging such as CECT or magnetic resonance imaging (MRI) is indicated to further characterize the cyst and assess its origin and extension. In this patient, CT scan was done as it was more readily available. Pre-operative

differential diagnosis based on the CT findings included exophytic, complex left liver lobe cyst, pancreatic pseudocyst, or splenic cyst. Pancreatic pseudocyst was considered less likely in the absence of previous trauma or episodes of pancreatitis and a normal looking pancreas. Complex liver cyst was favoured over splenic cyst in view of epidemiological prevalence of liver cyst in our local population and rarity of splenic cyst. Retrospective review of the CT images revealed the enhancing soft tissue at posterior aspect of cyst actually corresponds to the surrounding splenic parenchyma and was misinterpreted as thickened cyst wall. A thin sliver of compressed splenic parenchyma forming sharp interface with the cyst at its anterolateral and inferomedial aspect would further serve as a clue to the origin of the cyst arising from the spleen rather than left lobe of liver.

The definite diagnosis can only be made via histopathology. Most of the primary NPSC are solitary but rarely can be multiple. Histologically, NPSC have an epithelial lining and by using Immunohistochemistry the epithelial cells will be positive for pan cytokeratin and negative for CD 240 (lymphatic marker) and CD34 (an endothelial cell marker). In our case, the final pathological diagnosis was splenic epithelial (Mesothelial) cyst.

Total splenectomy has always been advocated in the treatment of symptomatic large cysts. Recently, spleen preserving surgical approach has been recommended due to the risk of overwhelming post-splenectomy infection (OPSI) (4%) and mortality rate (1.5%). Conservative methods include aspiration, drainage or injecting sclerosing agents but are associated with high recurrence rates and potential risk of bleeding and infection. Moreover, the inflammatory response can lead to dense adhesion and make subsequent surgery difficult. Surgical approach includes partial splenectomy, cystectomy, decapsulation and marsupialization via open or laparoscopic procedure. The preferred surgical method would be laparoscopic partial splenectomy if the cyst is small and done by a highly skilled and experienced laparoscopic surgeon. Cystectomy, decapsulation and marsupialization are simpler with minimal blood loss, but the disadvantaged is a higher recurrence rate. However, total splenectomy is indicated for giant splenic cyst occupying the whole spleen (like our case), or atrophic remnant spleen or splenic cyst involving the hilum.

CONCLUSION

Pre-operative diagnosis of giant NPSC remains a challenge and should be considered a differential diagnosis though rare. Laparoscopic and splenic preserving surgery is the preferred technique of treatment, but it is mainly for relatively small and peripherally located cyst. In our patient, we believe that total splenectomy was the best option and effective method to treat giant epithelial NPSC.

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DECLARATIONS

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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