CASE REPORT

First toddler mesenteric lymphatic malformation in Malaysia - A case report

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SUMMARY

This is the first reported case of a mesenteric lymphatic malformation in a toddler in Malaysia. It is a rare benign condition with incidence of 1 in 250,000 populations. Our patient presented to us at 2 years 11months old complaining of abdominal distension for 6 months without obstructive symptoms. Clinically there was a vague soft central abdominal mass. CT abdomen done revealed a large multiloculated intraperitoneal mesentery cystic mass within the central abdomen extending to pelvis. A semi-emergency laparotomy was performed. Intra-operatively the multiloculated mesenteric cyst measured 20cm x 30cm, adherent to the small bowel beginning at 12cm from duodeno-jejunal junction. Resection of the mesenteric cyst with adherent small bowel and primary anastomosis was done. Histopathological examination revealed multiple large lymphatic channels of various sizes in the mucosa and submucosa. Our patient has no signs of recurrence and remains symptom-free after 1 year since his surgery. Surgery with clear margins of resection is the recommended gold standard based on available literature. Type of surgical resection required will depend on the type of mesenteric lymphatic malformation. An awareness of this rare pathology is required to ensure proper management is given to these patients.

INTRODUCTION

Mesenteric lymphatic malformations are a rare benign condition which represent less than 1% of all lymphatic malformations. Its incidence is estimated to be 1 in 250,000 population.[1] To date this is only the third case of mesenteric lymphatic malformation reported in Malaysia, but the first one in toddlers.[2]

CASE REPORT

Our patient presented to the emergency department when he was 2 years 11 months baby boy with a complain of abdominal distension for 6 months but was able to have normal bowel openings without obstructive symptoms. Abdominal radiography performed showed no dilated bowels, however there was paucity of bowel gas centrally. On examination the abdomen appeared slightly distended with a central vaguely palpable soft mass. He was reviewed at the outpatient clinic and admitted. Sonography revealed a large intra-abdominal mixed solid cystic mass extending to pelvis with multiple thick septae and thick debris within the mass, likely lymphatic malformation of the mesentery. We proceeded with a CT of the abdomen (Figure 1) which showed a large multiloculated intraperitoneal mesentery cystic mass within the central abdomen extending to pelvis, multiple mesentery vessels insinuating the cystic lesion, no solid lesion with cystic mass, no bowel dilatation; likely mesenteric cyst. Alpha fetoprotein and beta-Hcg were normal. Patient underwent a semi-emergency midline laparotomy and excision of mesenteric cyst. Intra-operatively (Figure 2) there was a large multiloculated mesenteric cyst measuring 20cm x 30cm. The cyst was adherent to the small bowel beginning at 12cm from duodeno-jejunal junction. The 24cm of small bowel was resected along with the cyst with a remaining small bowel length of 211cm. Patient was discharged home well on post-operative day 5.

Histopathological examination (HPE) reported multiple cystic lesions at mesentery measuring 180mm x 70mm x 50mm with cysts sizes ranging from 5mm to 8mm. Sections from the bowel showed multiple large lymphatic channels of various sizes in the mucosa and predominantly in the submucosa. Similar cysts were also seen in the mesentery surrounded by loose connective tissue stroma. These cysts were lined by fairly uniform flattened to cuboidal epithelium and containing eosinophilic secretions. The lining epithelium were positive for CD31, focally for CD 34 and Factor VIII. They were negative for Calretinin. Focally disorganised smooth muscle are also noted within larger cyst wall. Lymphoid aggregates with some foreign body granuloma and cholesterol clefts were present at the periphery. No communication between the mesenteric cyst and the small intestinal segment was noted. No evidence of nuclear atypia or malignancy was seen. The surgical margins resected were adequate.

During the post-operative review at 1 month, 4 months and 1 year, the patient was well and did not have any complications or symptoms recurrence. No follow-up imaging was done.

DISCUSSION

Cystic mesenteric lymphatic malformations previously known as mesenteric lymphagiomas are divided into

This article was accepted: 18 October 2021
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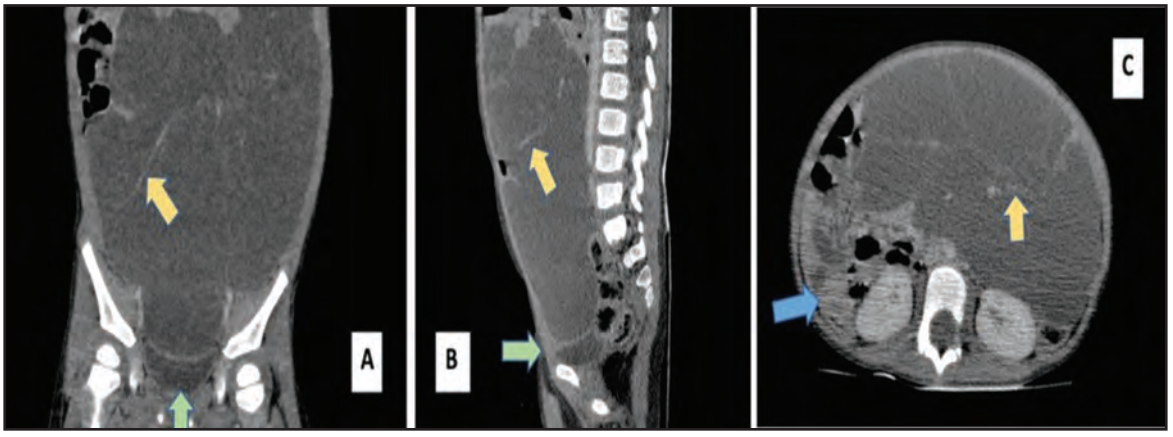

Fig. 1: Computerized tomography contrasted computed tomography (CT) scan images in coronal.
(a) sagittal (b) and axial (c) images shows large multiloculated intraperitoneal mesentery cystic mass within the abdomen (which is centrally located), below the subhepatic region and extends inferiorly until the pelvis (superior to the urinary bladder- green arrow). Bowel (blue arrow) is displaced postero- laterally by the large cystic mass. No extension of the mass into the spinal canal. Presence of multiple thin enhancing septa (yellow arrow) within the mass. No solid lesion seen within the cystic component.

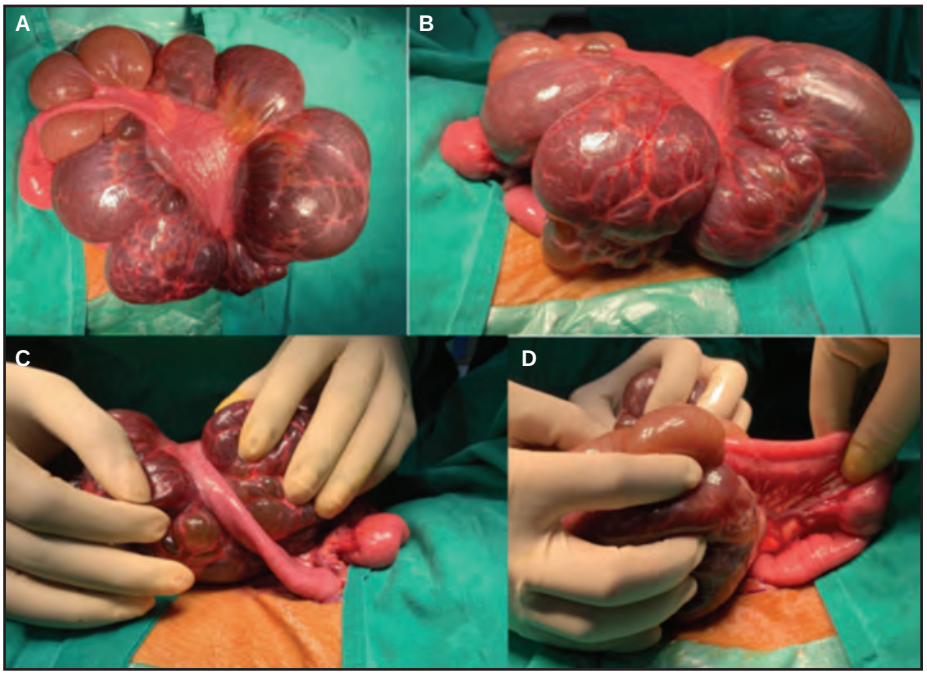

Fig. 2: Intra-operative findings revealed a large multiloculated mesenteric cystic lesion measuring 20cm x 30cm, a part of the small bowel adherent to cyst proximally 12cm from duodenojejunal junction. 24cm of small bowel was resected along with the cyst with a remaining small bowel length of 211cm. [Image A: Anteroposterior view. Image B: Lateral view. Image C: Inferior view. Image D: Caudal view.]

macrocystic, microcystic and mixed lymphatic malformations according to the latest ISSVA 2018 classification.[3] Lymphangiomas were previously classified histologically into capillary (simple), cavernous and cystic.[4]

The typical location for lymphatic malformations are head, neck and axillary region. Abdominal lymphatic malformations are rare and consist of only 5% of all lymphatic malformations, majority being diagnosed in childhood with 88% having symptoms such as abdominal distension, abdominal pain, abdominal mass, altered bowel habits, nausea or vomiting.[5] Abdominal lymphatic malformations may arise from the mesentery, solid organs (liver, spleen, pancreas), retroperitoneum and the gastrointestinal tract. Lymphatic malformations specifically of the small bowel mesentery has been described with an incidence of less than 1%.[4]

The aetiology of mesenteric lymphatic malformations were proposed to be due to congenital sequestration of lymphatic vessels during the embryonic period instead of a true lymphatic tumor. This is evidenced by majority of cases

presenting in childhood. In the older age groups, there are other etiologies proposed such as abdominal trauma, inflammation, radiation, abdominal surgery and lymphatic obstruction.[4]

In the literature there are two available gross anatomical classifications for mesenteric lymphatic malformations. The latest was proposed by Kim et al in 2016.[6] According to this classification, our patient would be of Type 1. The pathology in our patient is not clearly classified according to the previous classification proposed by Losanoff et al in 2003.[7]

Mesenteric lymphatic malformations appear as multiloculated cystic lesions on sonography, usually anechoic but may contain debris. Sometimes the lesions may appear as predominantly solid such as in our case where a solid component was seen on the ultrasound.[8] However, bearing in mind that ultrasounds are operator dependent, a CT scan or MRI will help to better delineate the anatomical structure of the malformation. The CT scan for our patient revealed a large multiloculated intraperitoneal mesentery cystic mass with no solid component.

The treatment for mesenteric lymphatic malformations are different from lymphatic malformations located in the common regions like head, neck and axilla. When originating from the mesentery, surgery with clear margins of resection remain the gold standard especially in symptomatic patients in which the lymphatic malformations has caused complications such as haemorrhagic infarction, intestinal obstruction, small bowel volvulus and infection. Surgery is also recommended even though the patient may be asymptomatic as there is a risk of progressive infiltration into retroperitoneal structures which will make future resection more difficult. The classifications of mesenteric lymphatic malformations are important to assist in surgical decision making of these patients. Based on the classification by Kim et al6, patients with Type 1 and 3 will need resection to include the bowel as blood supply will be affected. Those with Type 2 can have bowel sparing surgery as the malformation is pedicled and not affecting the blood supply of the bowel. Patients with Type 4 may not have adequate resected margins due to the extensive spread and may benefit from medical therapy such as propranolol, sirolimus or sclerotherapy.[1,6]

When mesenteric lymphatic malformations are located in the head, neck and axilla, surgery is avoided if the child is asymptomatic, not increasing in size and not affecting mobility or function. It is very important to restore and preserve the function and aesthetics of these regions. Other treatment modalities like OK-432, sirolimus, propranolol and sclerotherapy (using agents such as bleomycin, lauromacrogol, doxycycline, ethanol, etc.) would be the initial choice of therapy to reduce the size of these lymphatic malformations in order to achieve clear margins if surgical intervention is needed. Sclerotherapy is currently the gold standard of treatment for macrocystic or mixed lymphatic malformations in these regions.[9,10]

ACKNOWLEDGMENT

The authors would like to thank the Director General of Health Malaysia for the permission to publish this paper
NMRR ID: NMRR-21-1093-60075

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