



Mesenteric Cystic Lymphangioma “Intra-abdominal Catastrophe”

R. S. Kiran^{1*}, S. Sarmukh¹ and H. Azmi¹

¹*Department of General Surgery, Hospital Sultan Haji Ahmad Shah, Temerloh, Pahang, Malaysia.*

Authors' contributions

This work was carried out in collaboration among all authors. Author RSK is main and corresponding author for this manuscript. Author SS is responsible in supervising and editing of the manuscript and author HA is responsible in finalizing the manuscript. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Lymphangioma is a benign tumour of lymphatic origin. Lymphangioma in peritoneal cavity is extremely rare (5%), particularly in adults. Hereby, we are reporting a case of a Malay gentleman with no co morbidity presented with sign and symptoms of intestinal obstruction. We proceeded with exploratory laparotomy with small bowel resection with primary anastomosis. Intraoperatively, revealed soft mesenteric mass measuring around 5X5 cm and it was 80 cm from duodenum-jejunum flexure. Histopathological examination showed mesenteric cystic lymphangioma (MCL). Cystic intra-abdominal mass should be included MCL as a diagnosis although it's rare. Surgical management is still a mainstay of treatment due to its potential to invade vital structures and develop life threatening complication such as intestinal obstruction and bleeding. The surgical outcome after a complete clear margin resection can avoid recurrence.

Keywords: Lymphangioma; cystic; mesentery; intestinal obstruction; surgery.

1. INTRODUCTION

Mesenteric cystic lymphangioma (MCL) is a rare benign tumour of lymphatic origin. The later are encountered rarely in surgical practice, the frequency is less than 1 per 100,000 hospital admission [1]. Lymphangioma are usually located in head and neck (75%) and axilla (20%) in children [2]. MCL are commonly cystic and occur in the mesentery followed by omentum, mesocolon and retroperitoneum [3]. Aetiology is unclear, it may be associated with developmental anomalies of lymphatics or obstruction of lymphatic channels caused by bleeding or inflammation [4,5]. Pre-operative diagnosis is often difficult due to frequent silent course. MCL should be promptly managed as it may lead to life threatening condition.

2. CASE REPORT

A 61 year old Malay gentleman with no co morbidity presented with colic abdominal pain with no bowel output and reduced oral intake for three days duration. Upon assessment abdomen was distended with absent of bowel sound. There were no previous scars or mass, hernia orifice intact. Abdominal X-Ray showed features of small bowel obstruction (Fig. 1). He was subjected for exploratory laparotomy. Intraoperatively showed a soft mesenteric mass measuring around 5X5cm and 80cm from duodenum-jejunum flexure with adhesion to

pelvis causing transitional zone (Figs. 2 & 3). We performed segmental small bowel resection with primary end to end anastomosis. Intra-operatively, the rest of organ liver, gallbladder, spleen, stomach, pancreas and the rest of colon revealed no pathology. Post operatively patient had a speedy recovery and was discharged home well. During our outpatient clinic assessment (post-operative 2 weeks), patient was well with no new complain or symptoms, HPE reported as mesenteric cystic lymphangioma. We reassessed patient post-operative 4 months and discharged him as he was well.

3. DISCUSSION

Mesenteric cystic lymphangioma (MCL) are rarely encountered, fewer than 200 reports published in English language literature. In Malaysia we did encounter in other institution once presented as appendicular mass which turn out to be mesenteric lymphangioma. However, in my institution it was first case encountered [6]. Most commonly occur during childhood age and male predominant. Usually located in the neck (75%, also known as cystic hygroma), axilla (20%), less than 1% affect mesentery. Small bowel mesentery is frequent among intra-abdominal lymphangiomas. Other differential diagnosis includes pancreatic pseudocyst, echinococcal cyst, enteric duplication cyst and cystic mesothelioma. Classification of MCL as below (Tables 1 & 2).

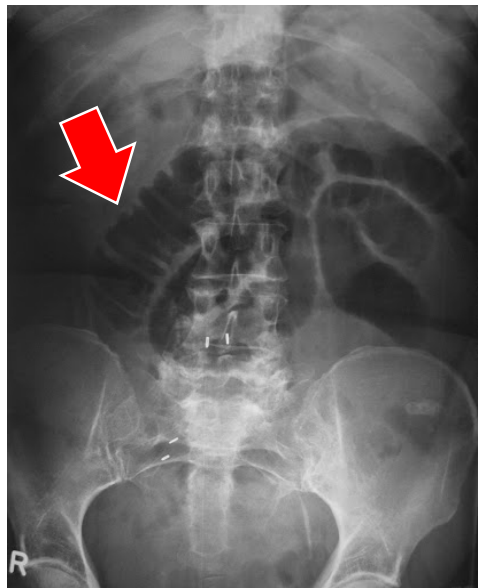


Fig. 1. Showing evidence of dilated small bowel on abdominal X-ray

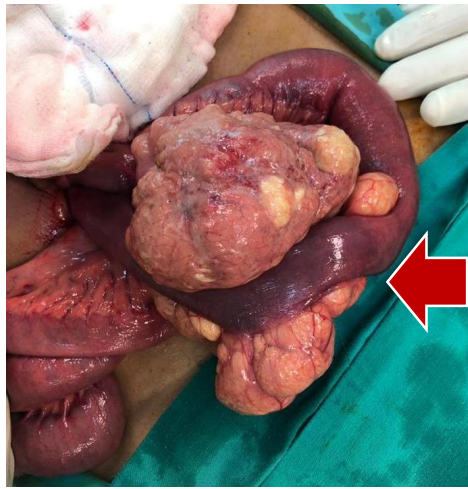


Fig. 2. Showing intra-operative finding of mesenteric cystic mass

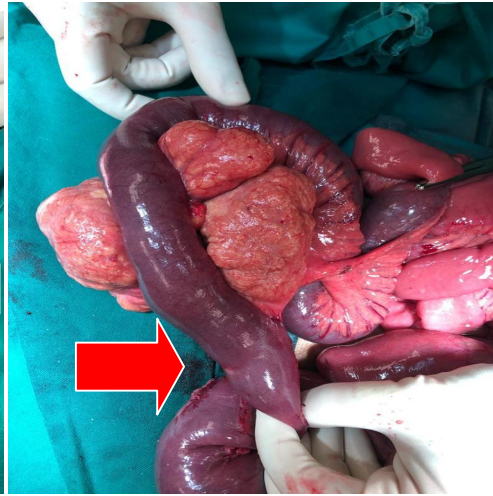


Fig. 3. Showing area of transitional zone

Table 1. Type of MCL [7,8,9,10,11]

Type	Description
Type 1 (Pedicled MCL)	Able to cause volvulus of intestine -> necrosis. Usually removed without risk of compromising intestinal blood supply.
Type 2 (Sessile MCL)	Located in mesentery boundaries and less mobile compare to type 1. Removal can affect bowel vascular supply hence requiring bowel resection.
Type 3 (Retroperitoneal extension)	Complete removal is impossible due to its involvement.
Type 4 (Multicentric)	Prognosis can be guarded if involves intra-abdominal and retroperitoneal organs.

Table 2. Classification of MCL [12]

Capillary	Predominantly cutaneous lesion
Cavernous	Predominantly cutaneous lesion
Cystic	Generally found in abdomen and retroperitoneum. Occurs in spaces surrounded by loose connective tissue such as mesentery and retroperitoneum.

MCL contents usually serous, serosanguinous or chylous fluid. Variability is due to degree of lymph stasis. Most clinical presentation vary from asymptomatic to abdominal symptoms to acute complication (such as volvulus, necrosis, rupture or bleeding).

However, definitive diagnosis is made by histopathology and immunochemistry. Histopathology examination will show cystic wall which are lined by flat endothelial cells and compromise of smooth muscle in the walls. Immunochemistry used for lymphangioma are Prox 1, CD 31, factor VIII related antigen and D2-40 which characterize the flat endothelial cells [13].

Mainstay of treatment for MCL is complete surgical excision of the mass. If there is infiltration of surrounding organ or involvement of main arterial branch, resection of involved organ would be required [14,15,16]. Other modality of treatment which could be offered such as drainage for high risk patients but successful rate is low as rate of recurrence is high and risk of mesentery perforation during the procedure is high [17]. Instillation sclerotherapy with alcohol can be used for ablation however it can cause normal tissue destruction [18]. In cases of remnant post excision, to prevent further enlargement of this remnants we could offer treatment (adjuvant) using OK-432 (biological response modifier with antitumor effects) [19].

Table 3. Showed the Diagnosis with aid of radiological tools [20,21,22]

Ultrasonography	For evaluation of abdominal cystic mass. Modality of choice for post-operative follow up. Described as a cystic lesion with thin septa (honeycomb or cobweb pattern)
Computed tomography (CT)	Provide information on location, surrounding organ involvement and size. It appears as a unilocular or multilocular mass with wall and septum enhancement by contrast. Is insufficient to establish an accurate diagnosis of MCL.
Magnetic resonance imaging (MRI)	As in CT aid in location, organ involvement and size. Most useful tool for establishment of MCL diagnosis. It clearly differentiates between cyst and lymphangioma in lack of fat content whereas in cyst they are clearly visualised in MRI.
^{99m}Tc-ASC (antimony sulfide colloid) lymphoscintigraphy	Provide lymphatic draining and anatomical information. In MCL it presents as multiple cystic lesions with increased tracer uptake in peripancreatic, retroperitoneal and iliac areas and abdominopelvic cavity. Is useful in diagnosing lymphangioma and demonstrate the communication between it and the lymphatic system. There was an observational study done on usefulness of this modality in diagnosing rare lymphatic disorder, one of it was lymphangioma (published in September 2020)

Prognosis and recurrent rate after complete surgical excision is considered to be excellent with almost completely no recurrence.

4. CONCLUSION

Mesenteric cystic lymphangioma (MCL) is a rare disease. Radiological assessment will aid in diagnosis preoperatively, however definitive diagnosis is made by histopathology and immunohistochemistry. Complete resection of tumour remains the mainstay treatment and to avoid recurrence.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist

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