A rare hypervascularised giant adult jejunal mesenteric lymphangioma

Aqlan Ahmad, MBBS[1], Wafaa Mohamad, MBBS[1], Kong Wai Keong, MD[1], Nil Amri MK, MD[1], Thanamogan Kerinon@Krishnan, MD[2]

[1]Department of Surgery, Hospital Sultanah Bahiyah, Alor Setar, Kedah, Malaysia, [2]Department of Pathology, Hospital Sultanah Bahiyah, Alor Setar, Kedah, Malaysia

SUMMARY

Mesenteric lymphangioma in an adult age group is a rare congenital benign tumour with an incidence of less than one percent. The tumour is biological slow growing. Thus, majority of patient present with non-specific symptoms and inconclusive imaging with mass effect which contributes to the difficulty in achieving the clinical diagnosis. Surgical excision of lymphangioma is the gold standard treatment but remains a surgical challenge due to the giant size intraabdominal lymphangioma. Here we present a rare case of an adult with an extreme sized mesenteric lymphangioma presenting with upper gastrointestinal symptoms. He successfully underwent surgical resection of the lesion which consists of 13 kg in weight and preservation of adjacent bowels.

INTRODUCTION

Mesenteric lymphangioma is an uncommon benign tumour with a reported incidence between 1 in 20,000–250,000.[1] Lymphangiomas are usually located in the head, neck, and axilla in children.[2] Almost 90% are detected by the mean age of two years.[3] Nonetheless, the risk of intraabdominal lymphangiomas which occur in adult age are extremely rare. Lymphangiomas occur at the mesentery in less than one percent of reported cases. The aetiology is unclear, but they are considered primarily to be congenital in origin.[3] Their suggested mechanism of occurrence is an anomalous development of the lymphatic system, which involves the obstruction of developed lymphatic channels due to lack of communication between small bowel lymphatic tissue and the main lymphatic vessels resulting in blind cystic lymphatic spaces. Most of the patients will present with non-specific gastrointestinal symptoms and with a huge sized mass effect obscuring a proper assessment view in imaging which makes it a challenge to make a clinical diagnosis. Surgical resection is the definitive treatment but remains a challenge in complete resection. Here, we present a case report of a giant size adult mesentery lymphangioma that underwent a successful complete resection with preservation of adjacent bowel.

CASE REPORT

A 29-year-old Malay gentleman presented to hospital with abdominal distension and early satiety for the past eight months. He had no vomiting or altered bowel habit to suggest of obstructive symptoms.

His body weight was 61kg. He was clinically thin in built with a grossly distended abdomen occupied by the mass which was non-tender and had the presence of a fluid thrill. Blood parameter showed no abnormality. Ultrasonography showed a large multiseptated cyst filling the whole abdomen. We proceeded with computed tomography (CT) of the abdomen and pelvis which showed a large abdominopelvic cystic mass (average HU of 20) with multiple enhancing septa mass effect to the surrounding structures and compression effect to the retroperitoneal urinary systems (Figure 1a and 1b).

Unfortunately, imaging was unable to determine the origin of the mass due to its large size. In view of its large size and multiple enhancing septa, it raised the suspicion of malignancy.

Tumour markers screening (alpha fetoprotein, carcinoembryonic antigen and CA 199) were normal. Infective screening for hepatitis B, C, HIV (human immunodeficiency virus) and VDRL (venereal disease research laboratory) was also not detected. He preceded with an ultrasound guided biopsy and cytology reported no malignant cells were seen.

He remained symptomatic and underwent laparotomy and tumour excision. Intraoperatively we found a large cystic lesion arising from the jejunal mesentery cyst occupying the entire abdomen. With the size of 41cm × 30 cm × 7cm. It was a hypervascularised lesion, well encapsulated, multilobulate, and multiseptated lesion (Figure 2a and 2b).

Initial working impression was of a mesenteric cyst.

The lesion was successfully excised en-bloc (Figure 2c and 2d) with preservation of major mesentery vessel and preservation of adjacent small bowel. Total cyst weight was 13kg. Colon, small bowel, rectum stomach, liver and spleen were normal with no peritoneal nodules.

Histopathology revealed the lesion was a mesenteric lymphangioma. Microscopically, the cystic lesion was composed of multiple large, irregular vascular spaces lined by flattened, single layered endothelial cells within a fibro-collagenous stroma, with no mitosis, atypia or multilayering of the lining cells seen (Figure 3a-d). In areas, the stroma shows infiltration of moderate lymphoplasmacytic cells and some reactive lymphoid follicle formation. Cholesterol clefts surrounded by foreign body type multinucleated giant cells

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Corresponding Author: Kong W.K
Email: kyouguang911@gmail.com

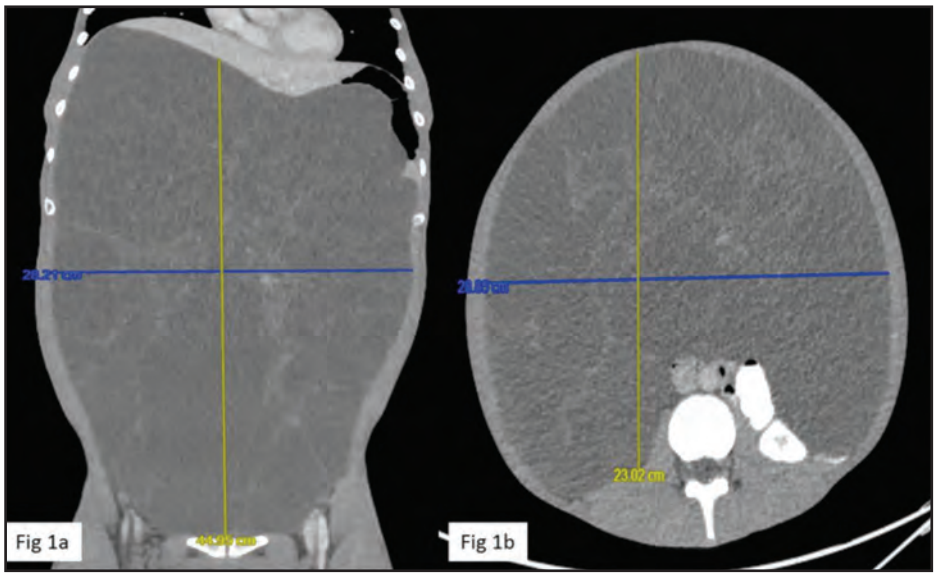

Fig. 1: (a) (CT coronal view)
(b) (CT axial view) showed huge multiple cystic enhancing lesion

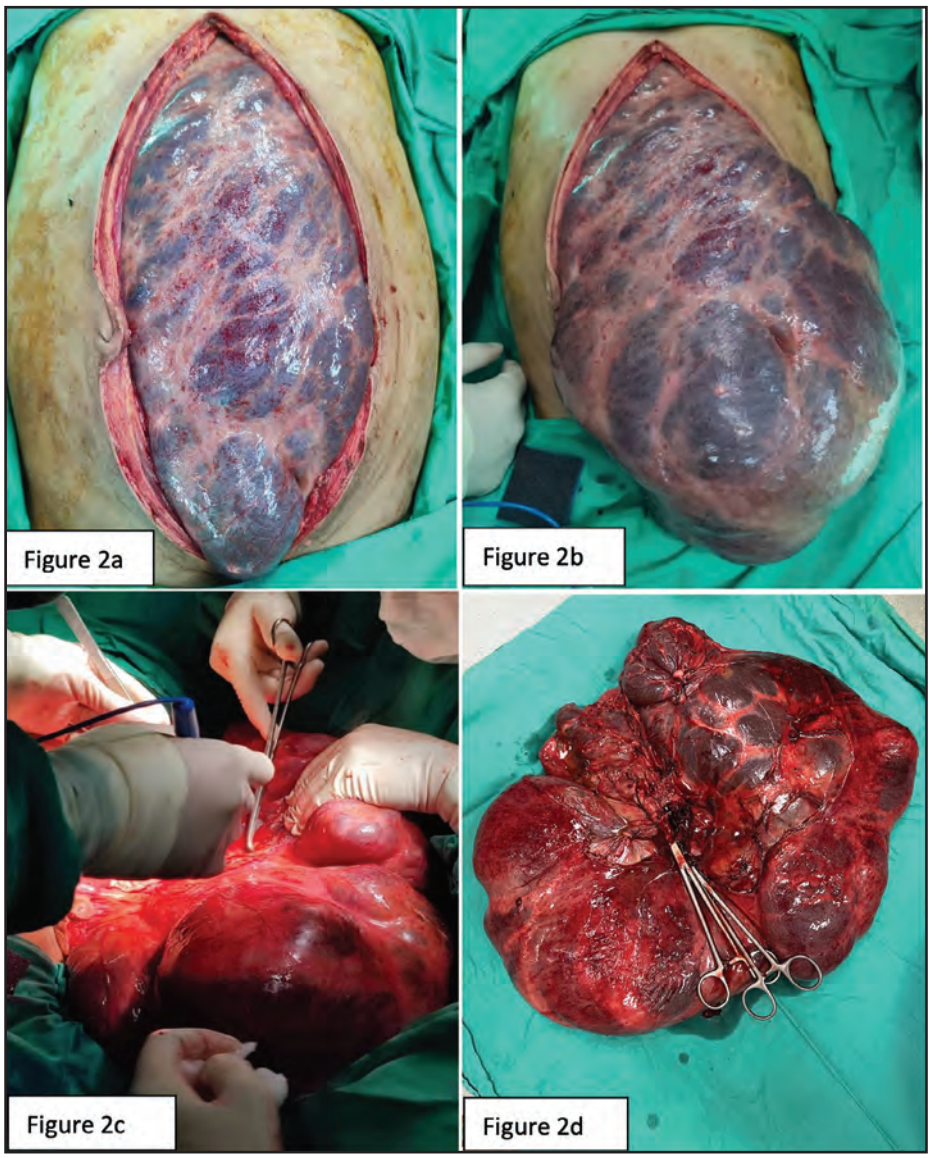

Fig. 2: (a) and (b) Intra-operative lesion prior excision
(c) Resecting the mesenteric lymphangioma
(d) Successful excised the mesenteric lymphangioma specimen

Table I: Classification of mesenteric cystic lymphangioma according to Losanoff and Kjossev

Types	Description et surgical possibilities
Type I	Pediculated with risk of torsion or volvulus. The resection is easy.
Type II	Sessile, less mobile that may require a nearby organ sacrifice
Type III	Includes a retroperitoneal extension (damage to vital structures sometimes) rendering total excision impossible
Type IV	Corresponds to extensive multi-organ damage

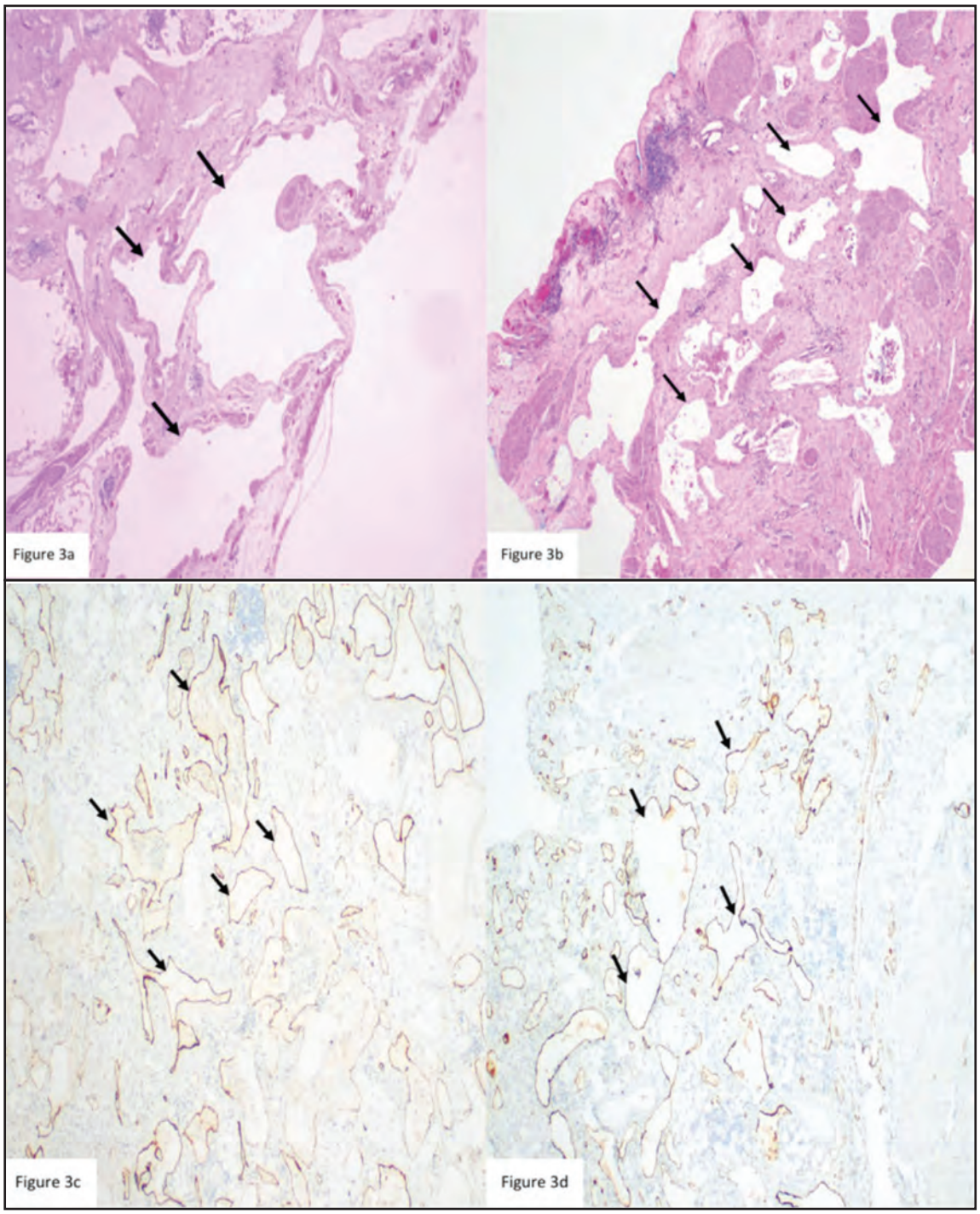

Fig. 3a & 3b : Cystic space lined by flattened, single layered endothelial cells (arrow)
(Figure 3a X20 magnification and Figure 3b X40 magnification)
Fig. 3c & 3d : D2-40(podoplanin) Immunohistochemical stain highlights the endothelial cells (arrow)

are also present. Smooth muscle bundles were seen in areas. No malignancy was identified.

DISCUSSION

Mesenteric lymphangioma are rare benign tumours which preoperative diagnosis is usually difficult due to the frequent silent clinical course. Patients may come with abdominal distension with or without intestinal obstruction and could present as incidental findings in asymptomatic patients.Some may present with acute abdomen such as rupture, infection, haemorrhage, or volvulus (4). Losanoff and Kjossev classification based on the morphotype of lesion is necessary to optimise the surgical treatment (5) (Table I). Radiological investigations are a useful diagnostic tool, but definitive diagnosis is confirmed by histopathology after a complete surgical resection.

Diagnosing an intra-abdominal cystic lesion needs thorough investigation to exclude malignancy and other differentials of an intra-abdominal cystic mass such as enteric duplication cysts, enteric cysts, mesothelial cysts, pancreatic pseudocysts, non-pancreatic pseudocysts, cystic mesotheliomas, cystic spindle cell tumours, and cystic teratomas. (6)

Ultrasound and CT are sensitive in providing structural details of the lesions. On the CT imaging, it appears as either uni- or multi-loculated configuration, size, enhancement of wall and septum.[2] However, ultrasound also can help to identify a cystic liquid lesion by its hypoechogenic and multiloculated morphology. Magnetic resonance imaging (MRI) is a more specific preoperative radiological tool for diagnosis and in surgical planning. It allows better differentiation of cystic and septal structures for comparison of mesenteric cyst and lymphangioma especially in mesenteric cystic lymphangioma. Mesenteric lymphangioma lacks demonstrable fat content by chemical shift and fat saturation.[7] In our patient, we did both CT and US imaging which clearly detailed the morphology and imaging characteristics of a cystic lymphangioma arising from the small bowel mesentery.

Accurate and definitive diagnosis is according to the histopathology and immunochemistry: the lining mesothelial cells are immunoreactive for cytokeratin and negative for factor VIIIIs. Staining with Prox1 and CD31 is the most reliable method for identifying lymphangioma endothelial cells.[8]

The main modality of treatment for mesenteric lymphangioma is adequate surgical excision, because there is a risk of recurrence and malignant transformation, particularly after radiotherapy for the primary lesion.[3] Lymphangiomas can become locally invasive and often require surgical excision, with reported recurrence rates of 12% and 53% when completely or partially resected, respectively.[9] Some cases require small bowel resection, but not in our case. It should be noted that the 10% of postoperative recurrence rate is due to incomplete resection, as evinced by positive microscopic resection margins. To prevent recurrence, complete excision of the tumour with or without intestinal resection is mandatory.

In our patient, we performed a complete surgical excision of the mass, that is the treatment of choice for cystic lymphangioma, even if asymptomatic. The prognosis after adequate excision of the cystic tumour of the mesentery was considered to be excellent.

Drainage has been suggested as a modality of treatment in high-risk patients but is often unsuccessful because of recurrence and for the risk of perforation of the mesentery during the drainage of the lymphangioma.[5] Instillation sclerotherapy with alcohol is being used for ablation, but this method can be destructive to normal tissue. In cases of failed percutaneous sclerotherapy using alcohol, acetic acid has been used with good success in intra-abdominal lymphangioma.[10]

Chen et. al in their study of six patients showed no recurrence during follow-up period of 3 to 12 months.[11] In our case, we follow up the patient three-monthly with a clinical examination and an abdominal ultrasound three months and six months after the operation which showed no recurrence.

CONCLUSION

Mesenteric lymphangioma are benign rare tumour in adults in which need thorough investigation. Surgical resection is gold standard and following up with imaging. Moreover, mesenteric lymphangioma in adult requires further study and clinicians should increase awareness to avoid misdiagnosis.

DECLARATION

The authors declare no conflict of interest with this work.

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