Unusual association of diseases/symptoms

Giant mesenteric lymphangioma: a rare cause of a life-threatening complication in an adult

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Summary

Mesenteric cyst lymphangiomas (MCLs) are rare benign tumours of unknown aetiology seen mostly in children. Clinical presentation can be diverse ranging from incidental abdominal cysts to an acute abdomen. A 24-year-old previously fit and healthy young man presented with a 2-week history of cramping, central abdominal pain and vomiting requiring acute hospital admission. He was a diagnostic conundrum despite extensive preoperative investigations. Recurring symptoms required urgent surgical exploration, resection and histological confirmation with a successful outcome. We review the literature and present here this rare case in an adult of MCL that has the potential to grow, invade and develop major life-threatening complications.

BACKGROUND

Mesenteric cyst lymphangiomas (MCLs) are commonly seen in children but rarely in adults, especially as a cause of recurrent bowel obstruction. Small bowel volvulus due to large MCL is very rarely reported in adults. Our patient's initial presentation was features of subacute bowel obstruction. Despite extensive investigations, recurrent volvulus of the small bowel due to the MLC was elusive. It was not until the surgical exploration that the cause of his recurrent symptoms was confirmed as MCL causing volvulus of the mid small bowel as a closed loop obstruction.

There is literature evidence that this rare tumour can present with life-threatening complications, including traumatic rupture, anaemia, intestinal gangrene secondary to volvulus and intermittent intestinal obstruction.

The reason for reporting this case is to emphasise the fact that the primary treatment of MCL is radical surgical excision, even when asymptomatic, because of its potential to grow enormously and invade adjacent structures, develop complications and its risk of sarcoma transformation on irradiation.

Our patient had a risk of life-threatening complication of ischaemia and infarction of the small bowel. Timely surgical intervention and radical excision helped to preserve most of the small bowel with a successful outcome.

CASE PRESENTATION

MCLs are rare benign tumours of unknown aetiology seen mostly in children. Clinical presentation can be diverse ranging from incidental abdominal cysts to acute abdomen.^{1 2}

A 24-year-old previously fit and healthy young man presented with a 2-week history of cramping, central abdominal pain, vomiting and distension requiring hospital admission. Past medical and family history was unremarkable. Physical examination revealed fullness and tenderness over the left lower abdomen. Fluid thrill and shifting dullness were negative.

INVESTIGATIONS

Blood and urine investigations were normal. Plain abdominal x-ray showed a large shadow displacing the left hemicolon with no features of bowel obstruction. The patient settled on conservative management. An ultrasound scan showed a multi-septate cystic lesion in the left abdomen and pelvis with a questionable origin. CT confirmed a large 20×16×15 cm multi-septate mixed solid and cystic lesion in the left flank and another 5×6 cm cystic lesion seen in the pelvis with a possible origin from the mesentery. We discussed the patient in the radiology meeting and cytological diagnosis was suggested. The 20 ml fluid aspirated following ultrasound demarcation was creamy in nature with no evidence of malignant cells on cytology.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included a multi-loculated mesenteric cyst.

TREATMENT

He was enlisted for an elective operative intervention. However, recurring symptoms of sub-acute bowel obstruction required an early exploration that showed a giant 24×20×15 cm thick-walled, multi-loculated mixed solid and cystic lesion with dark brown fluid originating in the mesentery and in close proximity and association with the jejunio-ileal junction. The entire mass was rotated 360 clockwise causing a closed loop obstruction of the mid small intestine. The mass was excised with clear margins, including 30 cm length of small bowel for vascular compromise (figures 1 and 2), and required a jejunio-ileal anastomosis for intestinal continuity.

Histopathological examination showed dilated lymphatics lined by attenuated endothelial cells confirmatory of cystic mesenteric lymphangioma.

Figure 1 The resected specimen (mesenteric cystic lymphangioma along with resected bowel).

Figure 2 Cut section of the mesenteric cystic lymphangioma.

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery and was discharged home on the fifth postoperative day. Follow-up at 8 weeks revealed he was fit and healthy and completely symptom free.

DISCUSSION

MCLs are rare benign tumours of the lymphatic system with a reported incidence of less than 1 per 100 000 hospital admissions.^{1 2} Exact aetiology is unknown. It is probably a developmental anomaly of the lymphatic system,³ explained by its primary occurrence in children, with over 80% diagnosed in the first few years of life.⁴ However, they can also form secondarily as a result of inflammatory process, lymphatic obstruction, surgery, radiation and abdominal trauma.^{5 6} Over 95% of lymphangiomas are found in the head, neck and axillary region. Isolated small bowel lesions are seen in less than 1% of cases,⁷ but this accounts for 70% of all intraperitoneal lymphatic tumours.

MCLs are classified as simple, cavernous and cystic.⁸ The simple type is mostly situated superficially in the skin and is composed of small thin-walled lymphatic vessels, while the cavernous variety has dilated lymphatic vessels and has connection with the normal adjacent lymphatics.⁸ Cystic lymphangioma, which was reported in our patient, is composed of large macroscopic lymphatic spaces surrounded by collagen and smooth muscles and does not have connection with adjacent normal lymphatics (CD34 and CD31 positive).⁸

Lymphangiomas are rarely seen in adults⁴ and are either discovered incidentally or may present with features of acute abdomen, as in our case, requiring urgent surgical intervention. There is literature evidence of this rare tumour presenting with life-threatening complications, which include traumatic rupture, anaemia secondary to intra-abdominal or intra-cavitary bleeding, intestinal gangrene secondary to volvulus and intermittent intestinal obstruction.^{1 5 8} Only three cases have been reported having some resemblance as noted in our case.

The role of plain abdominal film is debatable as no diagnostic features are confirmatory of lymphangiomas but would show evidence of intestinal obstruction.^{1 3} Ultrasonography, which has a high diagnostic yield for cystic lesions, is sensitive and relatively specific for diagnosing MCL.⁹ CT and MRI both give precise information of the size, anatomical location, extent of cysts and involved structures for the operative intervention;^{6 10} however, they are insufficient in establishing an accurate preoperative diagnosis⁹ as was evident in our case.

Primary treatment of MCL is radical surgical excision even when asymptomatic for its potential to grow enormously to invade adjacent structures, develop complications and its risk of sarcoma transformation on irradiation.^{6 10} However, radical excision can sometimes be technically impossible due to its infiltration of vital structures.¹¹ MCL has been classified into four categories by way of the relation between the cyst and the intra-abdominal anatomy (degree of mesenteric or abdominal organ involvement) to optimise surgical planning.^{1 6} Type 1: pedicled; type 2: sessile and less mobile and may alter the bowel's vascular supply requiring bowel resection, as in our case;³ type 3: retroperitoneal extension and can involve vital structures; and⁵

type 4: multicentric having doubtful prognosis especially in cases with extensive organ involvement.

Small bowel volvulus is rare in adults¹² with two distinct categories: primary type defined as torsion of the small bowel mesentery without evidence of predisposing anatomical abnormalities and secondary type precipitated by underlying anatomical abnormalities, including postoperative adhesion, malrotation, congenital bands, intussusceptions, colostomy, fistula, tumours and Meckel's diverticulum.¹² Large mesenteric masses can rarely cause volvulus of the small bowel.^{6 13 14} Our patient's initial presentation was features of subacute bowel obstruction. Despite extensive investigations, recurrent volvulus of the small bowel due to the MCL was elusive. He was enlisted for an elective exploration; however, recurrence of his symptoms required early intervention that confirmed the MCL causing volvulus of the mid small bowel as a closed loop obstruction accounting for his symptoms. Our patient had a risk of life-threatening complication of ischaemia and infarction of the small bowel. Timely surgical intervention and radical excision helped to preserve most of the small bowel with a successful outcome.

Our conclusion is that mesenteric lymphangioma should be included in the differential diagnosis of acute mechanical small bowel obstruction in a young adult in the absence of previous history of operative intervention and hernia.

Learning points

- ▶ Mesenteric lymphangioma should be included in the differential diagnosis of acute mechanical small bowel obstruction.
- ▶ Primary treatment of MCL is radical surgical excision even when asymptomatic because of its potential to invade adjacent structures.
- ▶ Preoperative investigations, including CT and MRI, give information about the size, anatomical location and extent of cysts; however, they are insufficient in establishing an accurate preoperative diagnosis.
- ▶ Early surgical exploration can prevent life-threatening complications that include traumatic rupture, intra-abdominal or intra-cavitary bleeding and intestinal gangrene secondary to volvulus.

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Competing interests None.

Patient consent Obtained.

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