CASE REPORT

Primary mesenteric lymphangioma in a young adult with intestinal malrotation and 'counter-clockwise barber pole sign'

Neha Singh, Ragini Singh, Udbhav Maheswari, Pallavi Aga

Department of Radiodiagnosis & Imaging, Chhatrapati Shahuji Maharaj Medical University, Lucknow, Uttar Pradesh, India

Correspondence to

Dr Neha Singh,
neha.singh.dr@gmail.com

SUMMARY

Mesenteric lymphangiomas are uncommon benign lesions that usually occur in isolation but rarely may be associated with gastrointestinal (GI) malrotation. Malrotation may cause chronic and recurrent volvulus leading to chronic venous congestion and lymphatic engorgement. Interference with lymphatic drainage may result in formation of a lymphangioma or a chylous mesenteric cyst. In our case, mesenteric lymphangioma was present with partial malrotation of small and large bowel. There was no radiological and surgical evidence of volvulus, favouring the hypothesis that lymphangioma may be a primary congenital association of GI malrotation. Another interesting finding was the counter-clockwise twisting of the superior mesenteric vein around the superior mesenteric artery which has been reported only seven times in the literature.

BACKGROUND

Lymphangioma, usually thought to be a paediatric disease, is a congenital malformation of the lymphatic vessels. Approximately 95% of lymphangiomas are found in the neck and axilla and the other 5% occur in the mediastinum and abdominal cavity, including the mesentery, retroperitoneum and bones.¹ Mesenteric lymphangiomas may produce complications such as intestinal obstruction, volvulus or infarction.² It may be primarily or secondarily associated with gastrointestinal (GI) malrotation.³ 'Counter-clockwise barberpole sign' was first described by Buranasiri *et al* as an angiographic sign in a rare case of malrotation with counter-clockwise volvulus.⁴ But studies have demonstrated that it may not always be associated with volvulus, rather it might be an association of GI malrotation or a normal variant.⁵

CASE PRESENTATION

A 22-year-old man presented with acute abdominal pain associated with a history of recurrent vomiting and obstipation for 1 month. Pain was episodic in nature (4–5 events in 24 h), colicky and of severe intensity. He was managed conservatively at a peripheral health centre for 3 days and was referred to Gastroscopy Department of King George's Medical University. The patient was pale and cachectic at the time of admission and had recurrent episodes of colicky abdominal pain and fullness. On examination, a soft, non-tender lump was palpable in the left hypochondrial and lumbar regions. Bowel sounds were hypoactive; both rest of the

abdomen and digital rectal examination were unremarkable. All blood and biochemical studies were within normal limits.

INVESTIGATIONS

Plain abdominal x-ray films demonstrated displacement of small bowel loops by a soft tissue mass towards the right side. Barium examination revealed abnormal location of duodeno-jejunal junction (DJJ). Apparently, the third part of the duodenum was located anterior to the spinous process of L2 (2nd lumbar) vertebra and the fourth part was looping back in a 'U' turn towards the right side, indicating malfixation of DJJ. Most of the jejunal and ileal loops were placed in the right half of the abdomen; only few small bowel loops were visualised in the left iliac fossa region (figure 1A). The caecum was visualised in the midline at the L3/L4 (3rd/4th lumbar) vertebral level, confirming the diagnosis of partial malrotation of the gut. On follow-up images, the ascending, transverse and descending colon was normally placed; however, the ascending colon was of very short length (figure 1B). An area of persistent non-filling was noted in the left hypochondrial and lumbar regions, which aroused the suspicion of an intra-abdominal mass, and ultrasonography (USG) was performed.

USG showed a multiloculated cystic lesion in the left side of the abdomen. Few vascular channels were noted to be traversing the lesion. The lesion was located medial to the spleen and anterior to the tail of the pancreas and the pelvic ileal loops, confirming its intraperitoneal location. USG findings were strongly suggestive of mesenteric lymphangioma (figure 2A). Another interesting finding was the anticlockwise twisting of the superior mesenteric vein (SMV) around the superior mesenteric artery (SMA) (figure 2B) with a mutual anatomical relationship maintained at the origin of SMA. For confirmation of the findings and characterisation of the lesion, multidetector CT (MDCT) was performed.

MDCT revealed a large well-defined cystic lesion of thin fluid attenuation in the left hypochondrial and lumbar regions. A mild enhancement of the cyst wall and septation was noted on contrast-enhanced scans. Encasement of branches of SMA and SMV was noted, confirming its origin from the small bowel mesentery. The lesion was extending up to the midline and causing a compression of the adjacent structures. Malposition of the DJJ and caecum with absence of the small bowel in the left side of the abdomen was also demonstrated.

To cite: Singh N, Singh R, Maheswari U, *et al*. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008994

Figure 1 Barium meal follow through showing: (A) the fourth part of the duodenum (D) looping back behind the third part of the duodenum (free running line). Duodeno-jejunal junction and jejunal loops (J) are bloated on the right side (black arrows) with a persistent filling defect on the left side. Stomach (St) is normally located. (B) Caecum (c) is placed towards the midline at the L3/4 level; ascending colon (As C) is of short length and passing towards the right side. Transverse colon (TC) is normally placed. Ap represents location of the appendix.

Counter-clockwise rotation of SMV around the SMA by 180–200° was noted to be approximately 3.5 cm distal to SMA origin known as the ‘Counter-clockwise Barber pole sign’; however, it was not associated with the twisting of the bowel or mesentery (figure 3). No sign of proximal bowel obstruction was noted. Based on the imaging findings, a final diagnosis of partial malrotation of the gut with mesenteric lymphangioma was made.

TREATMENT

After confirmation of diagnosis, the patient was kept nil orally for 24 h on intravenous fluids and through bowel preparation was made for surgery. At laparotomy a large cystic mass was identified occupying the whole left abdomen. The mass was mobile and lies anterior to the pancreatic tail in between the layers of the small bowel mesentery. On exploration DJJ was situated on the right side with the caecum shifted towards the midline confirming the preoperative diagnosis of malrotation. The mass was successfully excised in toto separate from the small bowel. Histopathological examination confirmed the diagnosis of mesenteric cystic lymphangioma (figure 4). Postoperative course was uneventful and the patient was discharged on the 15th day.

DISCUSSION

The estimated incidence of cystic lymphangiomas is approximately 1 per 20 000 paediatric population admitted in the hospitals.⁶ Sixty-five per cent of MCLs (mesenteric cystic

lymphangiomas) are congenital and 90% become symptomatic before the age of 2 years.⁷ The most common presentation is a palpable abdominal mass and abdominal distension.⁸ Intestinal obstruction related to the mesenteric cysts can be attributed to compression or traction by the mass.⁹ Although rare, a large mesenteric mass can rotate and cause volvulus of the connected mesentery and small bowel, and eventually closed loop obstruction of the involved bowel loops.¹⁰ Very few case reports describing cystic lymphangioma in adults are available in the published literature.¹¹

Different imaging modalities have been used to diagnose cystic intra-abdominal tumours. Some studies have demonstrated ultrasonography as being superior compared with CT regarding the assessment of intracystic structures such as the presence of echogenic contents, thickness of capsule and intracystic septation, which may indicate internal bleeding or infection. The typical CT feature of mesenteric lymphangioma is a multiloculated cystic mass with typically homogeneous fluid component of low attenuation values. Occasionally, negative attenuation values occur in the presence of chyle. Calcification may occur but is uncommon.¹² MDCT with thin slice multiplanar reformation is helpful not only to diagnose the lesion, but also to assess the complications such as volvulus and intestinal obstruction. MCLs that are relatively mobile have a tendency to give earlier complications and so are diagnosed sooner compared to retroperitoneal lymphangiomas. These complications include intra-abdominal bleeding, intestinal obstruction and bowel gangrene from volvulus, severe ascites and infarction of the cyst. Laparotomy may be necessary if signs of acute abdomen develop.

Pathogenesis remains unclear. The most accepted theory is based on a lymphatic congestion due to interrupted communication of the draining lymphatic system. However, this does not explain the occurrence of these cystic tumours in the adult population. Therefore, some authors believe that adult manifestations are the result of a delayed proliferation of congenital or acquired lymphoid nests after stimuli such as respiratory infection or local trauma.¹³

In our patient, partial intestinal malrotation was also present. In infants, the most common presentation is intestinal obstruction due to a mid-gut volvulus. In older children, the symptoms are usually atypical and vague, resulting in delayed diagnosis. The presentation is more frequently intermittent with episodes of spontaneously resolving bowel obstruction. This is thought to be due to kinking of the duodenum by Ladd bands rather than a volvulus. In patients with chronic and recurrent volvulus, chronic venous congestion with lymphatic engorgement may occur. Interference with lymphatic drainage may result in

Figure 2 Ultrasonography showing: (A) a multiloculated cystic lesion anterior to the pancreatic tail (1) and spleen (4). (2, 3 indicate splenic vessels). (B) Counter-clockwise twisting of the superior mesenteric vein (smv) around the superior mesenteric artery (sma).

Figure 3 Multidetector CT axial (A and B) and coronal (C) maximum intensity projection images showing: a large non-enhancing hypodense lesion (L) on the left side. The third part of the duodenum (D3) is passing anteriorly and the fourth part (D4) towards the right side. Counter-clockwise twisting of superior mesenteric vein (large arrow) around the superior mesenteric artery (small arrow) aka counter-clockwise barber pole sign is also seen.

formation of a lymphangioma.¹⁴ In our patient, counter-clockwise twisting of SMV around SMA was present without twisting of adjacent bowel loop or mesentery, excluding the possibility of volvulus. So, lymphangioma present in our case was not caused by volvulus, rather it was primarily associated with malrotation.

In total, there have been only seven reported radiological descriptions of counter-clockwise twisting of the SMV around the SMA known as 'counter-clockwise barber pole' sign. Of these, there were two rare cases of counter-clockwise volvulus,^{4 15} one case of malrotation without volvulus,⁵ one case of duodenal atresia with annular pancreas¹⁶ and three cases of suspected malrotation by upper GI series but normal rotation documented at surgery.¹⁷ In our case, twisting of vessels could be a primary congenital association of malrotation or a normal

anatomical variant as shown on MDCT and confirmed on surgical exploration.

Learning points

- ▶ In a young adult with recurrent abdominal pains, gastrointestinal malrotation should be considered in the differential diagnosis.
- ▶ Mesenteric lymphangioma may be primarily associated with malrotation or secondarily caused by concomitant volvulus.
- ▶ Twisting of superior mesenteric vein around superior mesenteric artery is not always associated with volvulus as it may be a normal anatomical variant or can be attributed to pressure effect of the cyst.
- ▶ Midline laparotomy and through abdominal exploration are mandatory in these cases to rule out volvulus and mesenteric hernias.

Figure 4 Histological section showing fibrous wall with adipose tissue (bare arrow) and cystic space filled with chylous fluid (black arrow) H&E ×100.

Contributors NS prepared the manuscript with the help of PA and UM under supervision of RS.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Lugo-Olivieri CH, Taylor GA. CT differentiation of large abdominal lymphangioma from ascites. *Pediatr Radiol* 1993;23:129–30.
- 2 Levy AD, Cantisani V, Miettinen M. Abdominal lymphangiomas: imaging features with pathologic correlation. *Am J Roentgenol* 2004;182:1485–91.
- 3 Antoniou D, Soutis M, Loukas I, et al. Mesenteric cystic lymphangioma associated with intestinal malrotation in a newborn. *Pediatr Med Chir* 2011;33:141–2.
- 4 Buranasiri SJ, Baum S, Nusbaum M, et al. The angiographic diagnosis of midgut malrotation with volvulus in adults. *Radiology* 1973;109:555–6.

- 5 Clark P, Ruess L. Counterclockwise barber-pole sign on CT: SMA/SMV variance without midgut malrotation. *Pediatr Radiol* 2005;35:1125–7.
- 6 Yildirim E, Dural K, Kaplan T, *et al*. Cystic lymphangioma: report of two atypical cases. *Interact Cardiovasc Thorac Surg* 2004;3:63–5.
- 7 Losanoff EJ, Richman BW, El-Sherif A, *et al*. Mesenteric cystic lymphangioma. *J Am Coll Surg* 2003;196:598–603.
- 8 Ros PR, Olmsted WW, Moser RP Jr, *et al*. Mesenteric and omental cysts: histologic classification with imaging correlation. *Radiology* 1987;164:327–32.
- 9 Mar CR, Pushpanathan C, Price D, *et al*. Best cases from the AFIP: omental lymphangioma with small-bowel volvulus. *Radiographics* 2003;23:847–51.
- 10 Al-Arfaj AA, Yaseen HA. Mesenteric cystic lymphangioma. *Saudi Med J* 2003;24:1130–2.
- 11 Losanoff JE, Kjossev KT. Mesenteric cystic lymphangioma: unusual cause of intra-abdominal catastrophe in an adult. *Int J Clin Pract* 2005;59:986–7.
- 12 Ko SF, Ng SH, Shieh CS, *et al*. Mesenteric cystic lymphangioma with myxoid degeneration: unusual CT and MR manifestations. *Pediatr Radiol* 1995;25:525–7.
- 13 Guinier D, Denué PO, Manton GA. Intra-abdominal cystic lymphangioma. *Am J Surg* 2006;191:706–7.
- 14 Yoon HK, Han BK. Chronic midgut volvulus with mesenteric lymphangioma: a case report. *Pediatr Radiol* 1998;28:611.
- 15 Pracros JP, Sann L, Genin G, *et al*. Ultrasound diagnosis of midgut volvulus: the 'whirlpool' sign. *Pediatr Radiol* 1992;22:18–20.
- 16 Hsu C, Chiba Y, Fukui O, *et al*. Counterclockwise barberpole sign on prenatal three-dimensional power Doppler sonography in a case of duodenal obstruction without intestinal malrotation. *J Clin Ultrasound* 2004;32:86–90.
- 17 Shimanuki Y, Aihara T, Takano H, *et al*. Clockwise whirlpool sign at color Doppler US: an objective and definite sign of midgut volvulus. *Radiology* 1996;199:261–4.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow