



Intermittent Small Bowel Volvulus Secondary to Mesenteric Lymphatic Malformation

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Abstract

Mesenteric Lymphatic Malformations (LM) are an unusual and yet important cause of abdominal mass in children and multi-modality imaging can support accurate diagnosis. We report the case of a four-month-old male who presented with symptoms of intermittent bowel obstruction. Radiographic imaging determined the presence of an intra-abdominal mass, displacing adjacent bowel. A combination of ultrasound and magnetic resonance imaging subsequently confirmed a multi-loculated, macrocystic structure of mesenteric origin with intra-cystic haemorrhage, consistent with mesenteric LM. When symptoms failed to respond to conservative management, definitive management involved surgical intervention. Intra-operative findings included a multi-cystic, lobulated lymphatic mass arising from the mesentery and encasing a segment of distal jejunum leading to deviation of the duodenal-jejunal junction to the right side. The mass was excised, along with the portion of encased jejunum and a wedge of mesentery. Final diagnosis was made of a mesenteric LM associated with intermittent localised small bowel volvulus secondary to encasement of jejunum and mass effect.

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Introduction

A four-month-old male presented with intermittent bilious vomiting, abdominal distension and progressive failure to thrive. We present the clinical history, pertinent imaging findings and outcome in this case of a macrocystic, mesenteric lymphatic malformation (LM) and intermittent volvulus. We will review the literature and discuss important aspects of differential diagnosis of mesenteric LMs in children.

Clinical history

A four-month-old male was taken to the Emergency Department of a District General Hospital due to multiple episodes of vomiting. He had vomited at least six times, including two episodes of bilious vomiting. Over the preceding four days, he

had been lethargic, not feeding well and had not opened his bowels. The baby had been born by normal vaginal delivery, at full term, without complication. He had passed meconium within the first twenty-four hours of birth. He had developed constipation at approximately five weeks of age, which had become more problematic in the three weeks prior to his presentation. He was born at 3.9 Kg and on the 75th centile, however his weight had gradually drifted down the centiles and he had failed to gain any weight during the previous month. On examination, he appeared irritable and slightly pale. His abdomen was mildly distended but soft, with no palpable masses. Rectal examination revealed a normally sited anus with normal faeces and no blood.



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Whilst initial observations and blood tests (including white cell count, C-reactive protein and serum lactate) were normal, an abdominal radiograph showed an abnormal gas pattern, with markedly dilated loops of bowel throughout the abdomen. He was transferred for assessment by the Paediatric Surgical Team at a specialist centre.

Imaging findings

Initial upper gastro-intestinal contrast study showed a normal position of the Duodenal-jejunal (DJ) flexure and no obstruction (Figure 1). Delayed follow-through showed a mass in the right flank, displacing the ascending colon medially, without radiographic evidence of acute bowel obstruction (Figure 2). An abdominal ultrasound was performed and demonstrated an 8 cm x 5.4 cm x 5 cm septated, avascular, cystic mass in the right flank, which was extending from below the right lobe of the liver into the pelvis and abutting the bladder. It was lying anterior to and separate from the normal right kidney, with its origin from the mesentery (Figure 3).

Subsequent Magnetic Resonance (MR) imaging confirmed the presence of this multi-loculated, cystic structure in the right flank. It returned low signal on T1-weighted and predominantly high signal on T2-weighted imaging. Fluid-fluid levels were seen in the cysts with some signal heterogeneity and areas of high signal on T1-weighted imaging, consistent with haemorrhage (Figure 4a & 4b). Radiological appearances were in-keeping with a right-sided LM with intra-lesional haemorrhage.

Differential diagnosis

Intra-abdominal LMs are uncommon and their radiological characteristics are not highly specific [1]. Differentiating LMs from other, more common, cystic masses in the abdomen, or from ascites, can pose a diagnostic challenge [2]. An important differential for a mesenteric cystic mass in a child is an enteric duplication cyst. Ultrasound evaluation is useful to visualise a multi-loculated mass with thin walls in the case of LMs, rather than the more multi-layered appearance of the enteric duplication cyst wall. If in continuity with the bowel wall, an enteric duplication cyst might also contain air, not seen in LMs. In female patients, an ovarian cyst ought to be considered in the differential diagnosis of cystic lesions – ultrasound is useful in identifying an ovarian origin [3].

On ultrasound, LMs are seen as multi-loculated, cystic structures, with echogenicity dependent on the subtype of LM. Macrocystic malformations show anechoic cysts with thin septations, in which internal echoes might represent haemorrhage, pus or chyle. Microcystic malformations show multiple solid lesions. Lesions are usually avascular, allowing differentiation from haemangiomas. Care should be taken not to interpret encased mesenteric vessels as vascularity [3]. MR imaging can be used to further characterise the lesion, by delineating the anatomical relations of the cystic mass and can also be particularly sensitive in identifying haemorrhagic complication. Typical signal pattern on MR resembles that of fluid, with low intensity signal on T1-weighted imaging and high intensity signal on T2-weighted images. Heterogeneity of signal can be seen secondary to blood, pus or chyle [1-3]. In this case, MR imaging established that the mass was not arising from the liver, spleen, pancreas or adrenal glands. The lesion appeared to originate from the mesentery and encase the jejunum. It also identified intra-lesional haemorrhage.

Outcome

The baby was initially managed conservatively and discharged when vomiting had resolved and feeding had improved. However, approximately one month later, he re-presented with recurrence of symptoms. A repeat upper gastro-intestinal contrast study again showed a normal position of the DJ flexure. The caecum was medially orientated and in a high position on a delayed radiograph, although on a subsequent radiograph it was more infero-medially positioned at the pelvic brim (Figure 5a & 5b). No features of obstruction were seen on the contrast study. A multidisciplinary decision was made for surgical intervention. Intra-operative findings were of a multi-cystic, lobulated lymphatic mass arising from the mesentery and encasing a 10 cm segment of distal jejunum (Figure 6a & 6b). Despite imaging showing a normal position of the DJ flexure, intra-operatively it was noted to be lying to the right of the midline and there was a highly mobile caecum, with a mildly narrow route of mesentery. The LM was excised with a wedge of mesentery and the distal jejunum it was encasing. An end-to-end anastomosis was formed and a limited Ladd's procedure was undertaken. He made a quick recovery and was discharged one week later.



Figure 1: Upper gastro-intestinal contrast study showing a normal position of the duodenal-jejunal (DJ) flexure and no obstruction.

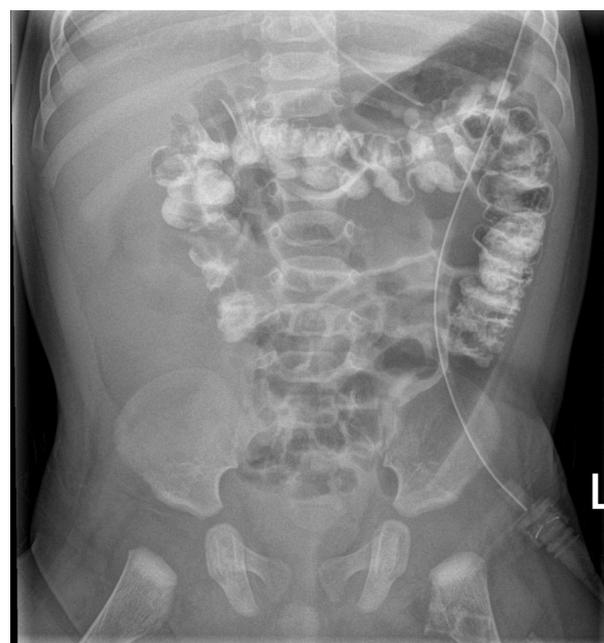


Figure 2: Delayed radiograph, showing a mass in the right flank, displacing the ascending colon medially.

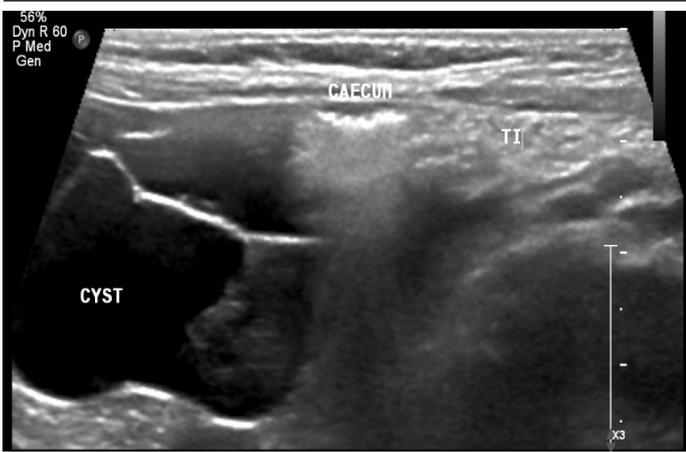


Figure 3: Abdominal ultrasound showing a septated, avascular, cystic mass in the right flank and its relationship to the caecum and terminal ileum (TI).



Figure 5: Delayed radiograph (at 2 hours post contrast administration via the nasogastric tube) showing the right flank mass, displacing the tortuous ascending colon superomedially and distal ileal loops medially. **(5a)**. Subsequent delayed radiograph (at 4.5 hours post contrast administration) showing a tortuous hepatic flexure, with the caecum inferiorly positioned **(5b)**.

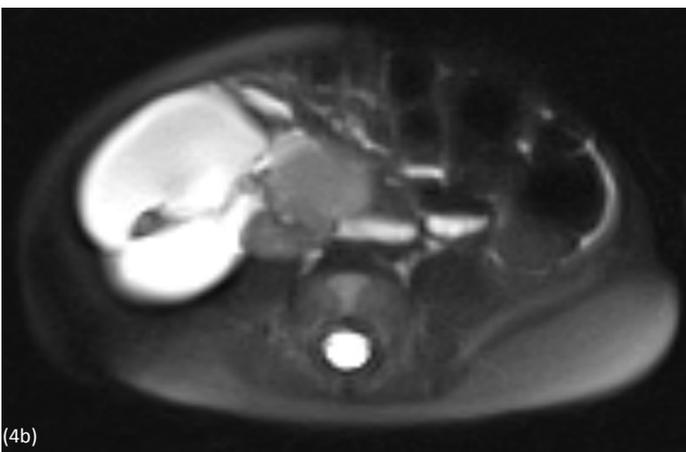
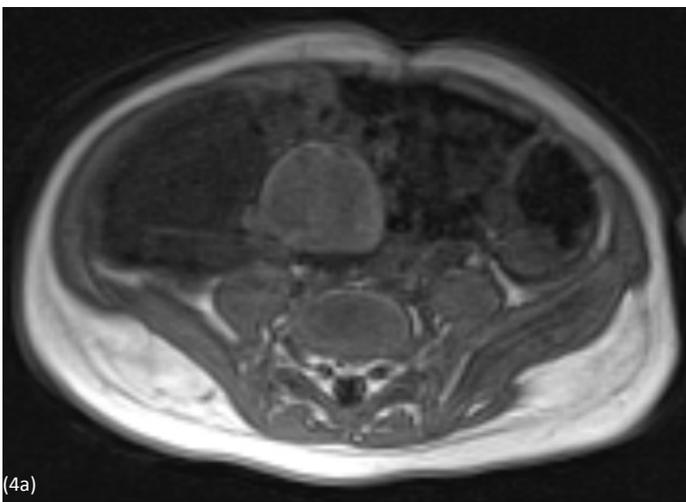


Figure 4: T1-weighted MRI showing a cystic structure in the right flank. Fluid-fluid levels are seen in the cysts with some signal heterogeneity and areas of high signal, consistent with haemorrhage **(4a)**. There is corresponding high signal on T2-weighted imaging **(4b)**.





Figure 6: Intra-operative findings included a multi-cystic lobulated lymphatic mass arising from the mesentery and encasing a segment of distal jejunum (6a), measuring 10 cm (6b).

Discussion

LMs are a type of simple vascular malformation that arise due to abnormal embryological development of the lymphatic system [4,5]. They most commonly present in childhood, with 90% presenting in children under two years of age [6]. They can occur almost anywhere in the body, however most often are located in the head and neck. Approximately 5% are found in the peritoneal cavity, retroperitoneum and mediastinum [2]. While they can be discovered incidentally, they can also present with symptoms due to mass effect on local structures. Intra-cystic haemorrhage, cyst torsion or rupture may also occur [7]. In this case, presentation was triggered by an intermittent localised small bowel volvulus due to the mass effect of the LM and encasement of the distal jejunum. Sonographic findings were in keeping with a LM and MR imaging was able to delineate the lesion and establish its origin from the mesentery. Surgical excision of symptomatic LMs is required when complications arise, which may involve, as in this case, resection of a portion of adjacent bowel [8].

Conclusion

Mesenteric lymphatic malformations are an unusual, yet important, cause of abdominal mass in children. Complications, including mass effect on adjacent structures, necessitate definitive surgical management and timely diagnosis, using multi-modality imaging, is fundamental.

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