A 10-year-old girl presented to the accident and emergency department of a public hospital with a 2-day history of vomiting, abdominal pain, and abdominal distension. She was admitted to hospital and a physical examination revealed a distended abdomen with sluggish bowel sounds. An abdominal X-ray showed distended small bowel in the left upper quadrant. Her blood tests were unremarkable. Initial management included fluid resuscitation, insertion of a nasogastric tube and a urinary catheter. Five hundred mL of bile-stained fluid was drained via the nasogastric tube. An emergency computed tomographic (CT) scan showed a distended small bowel and whirling of the superior mesenteric vein posterior to the superior mesenteric artery, suggestive of a volvulus (Fig 1).

A laparotomy was performed and revealed two cystic lesions over the jejunum causing a volvulus of the small bowel (Fig 2). The diseased jejunum was still viable, so the diseased small bowel was resected. The girl recovered well and was discharged 7 days after surgery.

An examination of the surgical specimens indicated that they were two interconnected cystic lesions measuring 7 x 3.5 x 2 cm and 3 x 2 x 2 cm over the serosal surface of the small bowel. The cysts contained 50 mL of brownish fluid and a microscopic examination showed that they were lined with regular endothelial cells. The cyst walls were made up of thin layers of smooth muscle and were supported by fibroadipose connective tissue. There were whitish plaques overlying the cysts, representing lymphangiectasia, consisting of dilated lymphatics in both the mucosal lamina propria and the submucosa. The pathology findings were compatible with lymphangioma with lymphangiectasia.

Lymphangiomas are rare tumours that usually occur in childhood. They are most commonly found in the neck and it is rare to find them sited intra-abdominally. Most intraperitoneal lymphangiomas are found in the mesentery, especially the small bowel mesentery. The aetiology is uncertain but a popular theory, the ‘blind sac’ theory hypothesis, postulates that a lack of lymphatic connections causes them to proliferate and dilate. This also explains why it usually occurs in the lymphatic-enriched mesentery of the small intestine. Although the lymphangiomas found in our patient were not located in the mesentery, the presence of lymphangiectasia supports the ‘blind sac’ theory. The cystic lymphangiomas may have developed from dilatation of an abnormal embryonic
Surgery is the treatment of choice for small bowel lymphangiomas, including mesenteric cystic lymphangiomas. Surgeons usually aim for complete removal of the tumour with surrounding organs of potential invasion, because there is a possibility of recurrence and invasion to surrounding organs. Partial or incomplete tumour removal may also be associated with complications like infection, fistula, and haemorrhage.

References