Presumed pancreatic cyst proven to be venous malformation due to intestinal malrotation

A 27-year-old man was admitted for evaluation of a 20-year history of chronic epigastric pain and weight loss of 3 kg over the preceding month. Despite several upper endoscopy examinations, no specific cause could be found. Physical examination showed mild epigastric tenderness. Laboratory findings were unremarkable apart from elevated creatinine level (3.17 mg/dL) due to focal segmental glomerulosclerosis. To investigate the cause of the abdominal pain, which became aggravated after a meal and improved with fasting, abdominal sonography was performed and showed a benign-looking 2-cm cystic lesion on the body of the pancreas (Fig. 1a). Although the cyst was devoid of intraluminal Doppler signal (Fig. 1b), it proved on abdominal magnetic resonance imaging (MRI) to be one of two focal venous aneurysms (Fig. 2a, b). Twisting of mesenteric vessels along the mesentery was also visualized on abdominal MRI (Fig. 2c, d).

Thus, both the focal venous aneurysms and the persistent abdominal pain could be attributable to intestinal malrotation. The findings of a small-bowel series were also compatible with intestinal malrotation: the distal duodenum and jejunum were arranged in a corkscrew appearance on the right side of the ligament of Treitz (Fig. 3a), and the cecum was also located in the right upper quadrant (Fig. 3b) [1]. Surgical management was planned, and on laparotomy the small intestine was found to be arranged in a coiled fashion (Fig. 4a). Ladd’s band could clearly be demonstrated and was excised to resolve the intestinal malrotation (Fig. 4b) [2].

Intestinal malrotation is a congenital anomaly of rotation of the intestine during embryonic development and occurs when the intestine does not make as many turns as it should [3]. It is most often recognized in the neonatal period and is rarely seen in adults. Since venous malformations due to intestinal malrotation can falsely manifest as pancreatic cysts, incidentally found pancreatic cysts in patients with abdominal pain should not be overlooked and additional imaging studies are necessary for further investigation.

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Fig. 3  Upper gastrointestinal series. a The distal duodenum and jejunum are arranged in a corkscrew appearance on the right side of the ligament of Treitz. b The cecum (arrow) is also located in the right upper quadrant.

Fig. 4  Ladd procedure. a The small intestine is arranged in a coiled fashion. b Ladd’s band (arrow). Excision of this band is the treatment for intestinal malrotation.

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