Recurrent acute pancreatitis in bowel malrotation

G. ALESSANDRI, A. AMODIO1, L. LANDONI1, N. DE’ LIGUORI CARINO, C. BASSI1

HPB Unit, Department of Surgery, Manchester Royal Infirmary Hospital, Central Manchester Foundation Trust, Manchester, UK
1Surgical Department, Pancreas Centre, Hospital of “G.B. Rossi”, University of Verona, Verona, Italy

Abstract. – OBJECTIVE: Recurrent acute pancreatitis is an uncommon diagnosis in teenagers. Excluded alcohol and biliary stones, more prevalent aetiologies in these group of patients are genetic pancreatitis, pancreatic duct system abnormalities, neoplasia, traumas and congenital abnormalities of the duodenum such as duodenal duplication or diverticulum. Two reported cases of recurrent pancreatitis associated to midgut malrotation were described in English literature. Bowel malrotation is a difficult diagnosis in a teenager or a young adult and a common delay is documented. Ladd’s procedure is largely recognized to be the standard treatment for a symptomatic malrotation.

CASE REPORT: Our Report describes in details a case of recurrent pancreatitis, where a late diagnosis of midgut malrotation was obtained and an endoscopic management was attempted. A literature review and an analysis of two previously reported cases were performed to explore a possible aetiopathogenesis of the recurrent acute pancreatitis in patients with midgut malrotation.

RESULTS: 24 months of follow-up showed an asymptomatic patient on a free diet, with a mild deranged Liver Functional Tests and a normal Amylase and Lipase. The recurrence of acute pancreatitis has not been observed until the present day.

CONCLUSIONS: Recurrent episodes of acute pancreatitis in young adults, without a history of alcohol abuse or evidence of gallstones, might be an atypical presentation of midgut malrotation and it should be in the differential diagnosis. In this case, a Ladd’s operation is beneficial and an endoscopic procedure does not obtain advantages.

Key Words: Recurrent acute pancreatitis, Uncommon diagnosis in young adult, Bowel malrotation, Ladd’s operation.

Introduction

Midgut malrotation is a not so rare congenital abnormality; occurring in one in 500 live birth1. The majority (approximately 80%)1,2 of these children develop gastrointestinal (GI) symptoms in the first month of their life. Midgut malrotation is described as a variation of the normal 270° counter clockwise rotation that starts after the fifth week of pregnancy. Generally, symptomatic patients present with an obstructive clinical picture; vomiting (generally bilious content), intermittent abdominal pain, diarrhoea, constipation, malabsorption and failure to thrive3. Midgut malrotation is always associated with an increased risk of bowel volvulus and a related ischic gut.

Pancreatitis is an atypical presentation of an abnormal intrauterine development of gastro-enteric apparatus. Pancreatitis during childhood is generally observed in children with genetic or congenital diseases causing cystic dilatation of bile ducts, maljunction of biliopancreatic ducts, pancreas divisum or other HPB malformation4-7. Causes of pancreatitis in an older age group (up to 25-30) are alcohol, CBD stones, trauma and congenital abnormalities of the duodenum such as duodenal duplication, intra or extra luminal diverticula. Rarely pancreatitis associated with midgut malrotation were documented in the literature; only two patients with recurrent episode of acute pancreatitis and a diagnosis of bowel malrotation were reported8-11.

Case Report

A 24-years-old woman was referred to our specialist Hepato-Biliary-Pancreatic (HPB) Centre for recurrent episodes of acute pancreatitis of...
unknown aetiology. Symptoms initially appeared in a fit and well patient 3 years previously; they were characterized by an epigastric and right upper quadrant colicky abdominal pain, very severe (VAS 10/10), radiated to the right back, with a gradual onset, associated with nausea and several episode of vomiting. On examination, the abdomen was tender in all upper quadrants with guarding on the right side but no rebound was estimated. First hospital assessment was in January 2009, blood showed mild deranged amylase (111 U/L) and Lipase (160 U/L) with normal Liver Functional Tests (LFTs). Three ultrasound scans (USS) could not show abnormalities. A CT scan (Figure 1) described an uncomplicated oedematous pancreatitis of body and tail of the gland and sludge within the gallbladder. The Common bile duct (CBD) was reported not dilated, the duodenum and the head of the pancreas were not described. A diagnosis of biliary pancreatitis was made and an elective laparoscopic cholecystectomy was performed after a full recovery from the episode of acute pancreatitis. The procedure was uneventful. Before the operation, a magnetic resonance cholangiopancreatography (MRCP) was obtained: “there were no gallstones, not dilatation of the CBD nor pancreatic duct” and there was not a further description of upper gastrointestinal (UGI) tract.

During the following year, the patient’s symptoms relapsed twice with a similar presentation; hospital admission and management with simple analgesia were required.

Eighteen months post-surgery recurrent episode of acute pancreatitis lead to a further admission. Investigations showed four times elevated amylase and lipase. Several abdominal and a pelvic ultrasounds were requested consecutively, no abnormalities were reported. A new MRCP estimated normal biliary tree and pancreatic duct, a CT scan with contrast showed no bowel obstruction but a “dysorphic head of the pancreas and a folded, festooned duodenum” were reported. An oesophago-gastro-duodenal endoscopy (OGD) was performed. It described “a considerable amount of biliary reflux into the stomach and normal first and second portion of duodenum”. An MRCP with secretin (Figure 2) was scheduled in a middle-high volume HPB centre to review the function of the Oddi sphincter. During this procedure, a bowel malrotation was described. The duodenum and the head of the pancreas were not lying on a retroperitoneal space, both of them were located between two layers of the peritoneum, a remnant of dorsal mesogastrium. The duodenum was twisted and mild retracted upward, the small bowel was situated to the right side of abdomen and the colon was on the left. The CBD, the main pancreatic duct (MPD) and the duodenum were anterior to the usual anatomy, more floppy and with an unnatural mobility. The pancreaticobiliary junction was described as normal. Neither of the CBD and the MPD were dilated, nor filling defects were appreciated and a normal pancreatic outflow was observed after the secretin stimulation. More investigations were requested and an endoscopic retrograde cholangiopancreatography (ERCP) showed: “a preapillary stricture of the main pancreatic duct without upstream dilatation”. A pancreatic sphincterotomy was performed during

Figure 1. The first CT scan performed on the patient showed the inverted relationship between superior mesenteric artery and vein, that was not reported.

Figure 2. MRCP with secretin revealed the uncommon presentation of the head of pancreas and the MPD (arrows). The CBD and MPD were not dilated, nor filling defects were appreciated and a normal pancreatic outflow were observed.
Recurrent acute pancreatitis in bowel malrotation

the ERCP and a plastic stent (4 cm, 5 Fr) left in situ for few days. A functional stricture and a bent duodenum were also confirmed.

After that procedure, the symptoms did not disappear and several episodes of recurrent pancreatitis were observed after that. Nine months later there was a new hospital admission. Amylase elevated at 6000 UI/L with mild deranged LFTs. USS and MRCP confirmed the uncomplicated pancreatitis and the ERCP showed a patent Wirsung and no stricture on the papilla. The patient was referred to our centre for recurrent pancreatitis.

Here a new assessment was obtained and all performed investigation reviewed. No mutation of CFTR and SPINK-1 were found on genetic tests. A contrast follows through (Figure 3) showed the bowel presentation. It was advocated to be responsible for a functional, intermittent obstruction of the duodenum. Thus, the increasing pressure inside the bowel leads to recurrence of symptoms. The mobility of the head of the pancreas was considered responsible for the functional obstruction to the pancreatic outlet. A Ladd’s procedure was scheduled and performed for the patient. A contrast follow through (Figure 4) was obtained one month after the procedure. No recurrence of symptoms has been observed after 24 months of follow-up.

Discussion

In literature, only two cases were reported with an association between midgut malrotation and recurrent episodes of acute pancreatitis. The first case described is a retrospective hypothesis that remains without objective proof.

The diagnosis of incomplete bowel rotation, Ladd ligament and an abnormal presentation of the duodenum were done only many years after the episodes of acute pancreatitis. At 9 and 10 years old the child had had two admissions with a diagnosis of idiopathic acute pancreatitis. Cholelithiasis, biochemical and infective aetiology had been excluded. Several USS had been performed at that time but a diagnosis of malrotation had not been obtained. The patient, at the age of 17, underwent to an exploration laparotomy for an acute intestinal obstruction. During the procedure, it was found a volvulus of the right colon secondary to a midgut malrotation. Authors just mentioned the attractive hypothesis of a recurrent acute pancreatitis due to a “distortion of the ampulla”, secondary to an uncommon presentation of the duodenum.

The second case is a 16 years old girl with a 6 years history of worsening recurrent acute pancreatitis. Several investigations were undertaken. A CT scan did not show abnormalities of the biliary tree, pancreas divisum or annular pancreas. A follow through study showed the presence of bowel malrotation and barium enema confirmed it. An ERCP was performed and it reported “a distortion of the first and second part of the duodenum associated with a mobile head of the pancreas. The CBD was mild dilated but the pancreatic duct was unremarkable and there was no abnormality of the pancreatic-biliary junction”. A laparoscopic Ladd’s procedure was performed eventually. There was no recurrence of symptoms during the 3 years following the procedure.
Sasaki et al.\textsuperscript{11} probed a possible aetiology with several investigation. Symptoms were related to an unusual feature of the duodenum and pancreatic head, twisted and retracted upward. This unusual UGI presentation was assumed to be responsible for a functional, intermittent pancreatic duct obstruction and consequently recurrent pancreatitis. Findings obtained in that patient by a follow through study and an ERCP supported the hypothesis. The lack of recurrence of symptoms during the following 3 years of follow-up supported authors opinion.

It is well documented as an inflammation of the pancreas can be obtained in an experimental way on mice, blocking the proximal part of the small bowel, distally to the papilla of Vater and before it, increasing the intraduodenal local pressure\textsuperscript{12}.

The non-specific presentation of our patient causes difficulty in establishing a diagnosis. After the first episode of abdominal pain, she was managed as a biliary pancreatitis\textsuperscript{13}. A conservative management of pancreatitis and an elective laparoscopic cholecystectomy were performed\textsuperscript{14,15}. Recurrence of symptoms were carefully investigated by MRCP, CT abdomen, USS and a MRCP with secretin (Figure 2). Eventually, an ERCP was scheduled. It reported: “a prepyloric stricture of the main pancreatic duct without upstream dilatation and normal outflow, in a patient with bowel malrotation”. According to this diagnosis, a pancreatic sphincterotom y was performed, but recurrences were observed again. When the patient arrived in our centre, a simple UGI series (Figure 3) was requested and the case discussed in a multi-disciplinary meeting. All available images were reassessed. A midgut malrotation with a Ladd’s ligament was confirmed and judged to be responsible for recurrent pancreatitis. Intraoperative findings supported our impression. In patient assessed in our centre, there were no signs of biliary obstruction. There was no abnormality in the main pancreatic duct, that was no dilated and without any irregularity of the wall. The unusual presentation of the duodenum and the small bowel were remarkable. The duodenojejunal junction was not found in the conventional position. The duodenum had an irregular twisted course, located in the right upper quadrant of the abdomen and with a Z shape (Figure 3). Peritoneal bands entrapped the descending and transverse portion of the duodenum (Ladd’s bands), responsible for its fixity to the retroperitoneal space, showing a variable degree of bowel obstruction. The pancreatic head and the latter part of the duodenum were anterior to the retroperitoneal plan, with a degree of mobility. This increased the possibility of an obstruction of the MPD as reported by Sasaki et al\textsuperscript{11}. The duodenum was closely related to a malrotated cecum and ascending colon. They were attached to the right side of the abdomen by peritoneal bands (Ladd’s ligament) including part of the duodenum. In this patient a pancreatic sphincterotomy and a stent did not settle the symptoms, showing that pancreatic duct obstruction was not related to a “distortion of the Ampulla of Vater”, as mentioned by Kirby et al.\textsuperscript{8} The particular presentation of the proximal midgut instead might be advocated as responsible of a variable duodenal obstruction. This type of bowel obstruction, with an intermittent kinking of the pancreatic duct, might be considered in our patient as a possible contributive aetiopathogenic mechanism of the recurrent acute pancreatitis.

In all these cases there was a delay in the diagnosis of midgut malrotation. Spigland et al.\textsuperscript{10} reported generally the delay of 20 months to reach the diagnosis of malrotation in teenager. In our case, symptoms initially started in January 2009 and the abnormal bowel presentation was detected in September 2010. Numerous USS, MRI and CT scans were performed.

Reassessing all available imaging of our patient obtained before the diagnosis, it was possible to observe typical features of incomplete rotation of gut. US, CT and MRI are not the preferred modalities to make a diagnosis of malrotation. Careful evaluation of all bowel and the inverted relationship between superior mesenteric artery and vein\textsuperscript{16,18} (Figure 1) can help to understand the anatomy. The gold start investigation to assess the first part of the GI tract is the contrast follow through study. There are three main radiological signs to detect an UGI malrotation: (1) abnormal position of the duodenojejunal junction; (2) spiral, “corkscrew” or Z-shaped course of duodenum and proximal jejunum; (3) a jejunum located in the right quadrants. In some patients, a contrast enema might be useful to confirm the diagnosis, as it can show the large bowel located on the left side of the abdomen.

In our case and in the case described by Sasaki et al, the Ladd’s operation was performed electively, which is widely recognized as the procedure of choice to correct a bowel malrotation. Several authors advocate a surgical correction in all patient with a diagnosis of malformation\textsuperscript{19,20}, regardless the presence of symptoms. A significant percent-
age of patients dies or suffers substantial morbidity due to loss of gut, secondary to a complicating volvulus and ischemic gut\textsuperscript{1-21}. Asymptomatic patients with a bowel malrotation will always have an increased risk of volvulus\textsuperscript{22-23}.

Our patient underwent an endoscopic pancreatic sphincterotomy before surgery. Recurrence of symptoms followed in several occasions. In our institute, an open Ladd’s procedure was performed. During the laparotomy there was a confirmation of the diagnosis. Ladd’s band was incised, the duodenum was carefully mobilized and manipulated straight. A duodenopexy was created and the duodenum was relocated to the right side of the abdomen in a non-twisted position, as described by Bax and Van der Zee\textsuperscript{24}. The appendectomy was not conducted and broadening of the mesentery was deemed not necessary\textsuperscript{25,26}. The surgical procedure was based on intraoperative finding; a “complete not rotation” pattern\textsuperscript{27}, the cecum located on the right lower quadrant and a general intra-abdominal assessment suggested a broad mesentery with a low risk for complicating volvulus after duodenopexy\textsuperscript{22,23}.

**Conclusions**

Congenital abnormalities have to be considered as a possible etiology of recurrent acute pancreatitis in a teenager, when the abuse of alcohol and the diagnosis of gallstones are excluded. The gold standard to make a diagnosis of midgut malrotation remain the gastrografin follow through study. However, a careful assessment of the presentation of the small bowel and the relationship between the mesenteric vessels can be used in all other cross section studies to obtain the diagnosis. A symptomatic patient with midgut malrotation needs a Ladd’s procedure. Recurrent acute pancreatitis can be included in the clinical picture of a symptomatic patient with midgut malrotation.

**Conflict of Interest**

The Authors declare that they have no conflict of interests.

**References**


26) **Ladd WE, Gross RE.** Intestinal obstruction resulting from malrotation of the intestines and colon in abdominal surgery of infancy and childhood, Chap 5. WB Saunders, Philadelphia, 1941; pp. 53-70.