Ectopic varices and superior mesenteric vein thrombosis as a result of midgut volvulus

N. Sikas¹, N. Nikolaidis², V. Papanikolaou¹, Olga Giouleme², D. Gakis¹, N. Evgenidis², A. Antoniadis¹

Summary
Midgut volvulus as a result of malrotation classically presents in early life and is associated with intestinal obstruction and compromised vascular supply of the gut. On rare occasions this condition may manifest itself in teenagers and adults, often with disastrous outcome. We report a 28-year-old patient who presented with melaena and endoscopy showing varices in the duodenum and large bowel. During the neonatal period of his life he had been operated on for intestinal obstruction, but the details of the operation were unknown to us. Angiography demonstrated superior mesenteric vein thrombosis and ectopic varices. On laparotomy he was found to have midgut volvulus. Recommendations for diagnosis and treatment are discussed.

Introduction
Midgut volvulus results in twisting of the intestine, causing intestinal obstruction and occlusion of the mesenteric vessels.¹ Usually, it presents in early life with symptoms of mechanical ileus or bowel ischaemia. In cases where the twisting is mild and has a progressive course, it causes gradual occlusion of the mesenteric vessels.² The superior mesenteric vein (SMV) collapses first and collateral circulation develops, draining the blood from the intestine. In this way colon, small bowel and duodenum varices arise.

Varices in the gastro-intestinal (GI) tract most commonly occur in the gastro-oesophageal region.³ Extra-oesophageal or ectopic varices can arise at other sites, such as duodenum,⁴ small bowel,⁵ colon⁶ and rectum.⁷ Portal hypertension is the most common cause of varice formation.⁸

We describe a case of an adult patient whose malrotation had not been diagnosed in early life and resulted in SMV thrombosis and ectopic varices. Review of the literature and the surgical management are discussed.

Case Report
A 28-year-old male patient presented with a 3-week history of giddiness. He denied any nausea, vomiting, haematemesis, haematochezia, recent weight loss or change in bowel movements. From the past medical history it was found that 28 years earlier, when he was 7 days old, he had undergone an urgent exploratory laparotomy for persistent bilious vomiting. The only information we could obtain had been that his condition was very serious, he had been critically ill and underwent gastrointestinal anastomosis, followed by an uneventful recovery. Physical examination revealed pallor, tachycardia and orthostatic hypotension while rectal examination showed black-colored, guaiac-positive stool. Laboratory values included haemoglobin of 5.9 mg/dl, haematocrit of 16%, platelets of 233,000 and prothrombin time of 13 seconds with a partial thromboplastine time of 29 seconds. His biochemistry, liver function tests, urinalysis and clotting screen were all within normal limits.

After adequate resuscitation with fluids and blood replacement the patient underwent oesophago-gastro-duodenoscopy (OGD). The oesophagus was normal with no evidence of varices. The fundus, body and antrum of the stomach had no abnormality. A gastro-intestinal anas-

¹Transplantation Unit & ²2nd Propaedeutic Dept. of Internal Medicine, Hippokration General Hospital, Thessaloniki, Greece

Author for correspondence:
Nicholas Sikas, 29A Martiou str., N. 751, Panorama, 552 36 Thessaloniki, Greece, Tel.: (+30-31) 341606, (+30-977) 832641, Fax: (+30-31) 855566, e-mail: niksik63@otenet.gr
tomosis was noticed. In the second part of the duode-
um large varices were found with no signs of active bleeding (Fig. 1). Colonoscopy showed a normal muco-
sa with extensive varices along the ascending and trans-
verse colon (Fig. 2). A digital subtraction angiography fol-
lowed. This demonstrated absence of the SMV, ex-
tensive collateral veins, draining the blood from the in-
testine into the splenic and portal vein mainly through
an enlarged gastro-duodenal vein and distended me-
senteric veins (Fig. 3, 4). Computed tomography of the
abdomen did not show any significant finding.

In view of the patient’s symptoms and in an effort to
establish the cause of the SMV thrombosis surgical in-
tervention was recommended. Exploratory laparotomy
showed a caecum that was located in the left hypochon-

Fig. 1. Gastroscopy showing varices in the second part of the
duodenum (arrow).

Fig. 2. Colonoscopy demonstrating a prominent varix (arrow).

Fig. 3. Digital subtraction angiography (DSA) showing absence of
SMV and a distended gastro-duodenal vein.

Fig. 4. DSA showing prominent mesenteric veins to the right
of the spine.


drium, while the whole ascending colon with the hepatic
flexure was mobile. The adhesions from the previous
operation were localized and by no means could have
interfered with the above finding. There was volvulus of
the mesentery that was reduced and the base of the me-
sentery was revealed together with the duodenum. Since
there was no need for the gastro-intestinal anastomosis,
which had apparently been performed 28 years earlier
to treat the duodenal obstruction, this was closed and
the continuity of the GI tract was restored. The caecum
and ascending colon were sutured to the right iliac fos-
sa. A peripheral branch of the SMV was anastomosed,
side to side, to inferior vena cava (Fig. 5). The pressure
in the SMV prior to anastomosis was 21 cmH\textsubscript{2}O (nor-
mally between 10-15 cm$H_2O$) and following the anastomo-
sis was 14 cm$H_2O$. The patient made an untoward
recovery, and one year following the operation he re-
mains asymptomatic and repeat gastroscopy and colon-
oscopy have not shown any varices.

**DISCUSSION**

Midgut volvulus is one of the abnormalities of mal-
rotation. In this condition the small bowel fails to enter
the abdominal cavity normally. As a result, the mesenter-
ies fail to undergo normal fixation and both the loops of
small intestine and the superior mesenteric vessels are
suspended from a narrow pedicle. Peristalsis may pro-
duce a torque in the loops, and the pedicle may twist,
resulting in intestinal obstruction or compromised me-
senteric vascular supply.

It usually presents in neonatal and paediatric popu-
lations. In this age group, most of the patients are younger
than eight weeks of age. The most predominant symp-
toms are bilious vomiting and constipation. Later as is-
chaemia advances, gangrene of the bowel occurs, asso-
ciated with high mortality. Occasionally though, midgut
volvulus may manifest itself in teenagers and adults,
sometimes with a disastrous outcome. Most frequently,
these patients will present with chronic abdominal pain,
with or without vomiting, or chronic diarrhea. Also they
may present with severe abdominal pain having inconsis-
tent clinical findings or atypical presentation of small
bowel obstruction.

On very rare occasions, gradual occlusion of the su-
perior mesenteric vessels secondary to recurrent or per-
sistent twisting of the mesentery, may result in occlusion
of the main vessels and development of arterial and ve-
nous collateral circulation. The later present with ec-
topic varices and recurrent gastro-intestinal bleeding. In
our patient only the SMV was occluded, presumably be-
cause the twisting was not severe enough to occlude the
superior mesenteric artery as well.

The diagnostic methods vary, depending on the clin-
cal presentation. In neonates and children, abdominal
roentgenograms with or without contrast, may demon-
strate the malrotation. For sick infants with signs of peri-
tonitis, contrast studies are not indicated. Exploratory
laparotomy is the only choice before ischaemia becomes
irreversible. Screening with ultrasound scan in patients
with suspected pyloric stenosis or acute abdomen plays
a significant role in the differential diagnosis. Findings
like fluid-filled, distended duodenum, dilated and thick-
walled bowel loops to the right of the spine and assess-
ment of the relative positions of the mesenteric vessels

![Fig. 5. Isolation of the Inferior Vena Cava and peripheral branch of the Superior Mesenteric Vein (A). Side-to-side anastomosis between the two vessels (B).](image-url)
should lead the sonologist to suspect midgut malrotation complicated by volvulus. One case has been reported where intestinal malrotation was an incidental finding on computed tomography.

Apart from the previously mentioned diagnostic modalities, one should also consider angiography and endoscopy of the GI tract, especially in those patients who present with episodes of gastro-intestinal bleeding. Angiography will demonstrate the absence of the superior mesenteric artery (SMA) or SMV or both, development of collateral veins, branches of the superior mesenteric vessels to the right of the spine and the characteristic “barber pole sign”. Other causes of chronic occlusion mesenteric vessels, such as pseudocyst, pancreatic or retroperitoneal neoplasms (lymphoma), Crohn’s disease, coagulopathies (polycythemia vera, protein C or S deficiencies), sickle-cell anemia, contraceptives, portal hypertension and idiopathic SMV absence, should be excluded.

Endoscopy will reveal ectopic varices, usually in the duodenum representing enlargement of the gastroduodenal vein branches. Also ectopic varices will be found in the small bowel, ascending and transverse colon. These represent enlarged mesenteric veins secondary to venous stasis.

For ectopic varices located in the colon, colonoscopy is of great help, bearing in mind that its sensitivity is greatly reduced during periods of active bleeding and when adequate mechanical bowel preparation has not been achieved. Increased intraluminal pressure due to air insufflation may result in failure to identify these lesions. The varices appear as tortuous vascular dilations with a bluish discoloration or they may resemble polyps and, if biopsied, will bleed profusely.

In the recent years laparoscopy has become an excellent technique for the evaluation and definite management of patients with intestinal rotation abnormalities.

The most common cause of varices in the GI tract is portal hypertension. These varices are usually located in the gastro-oesophageal region where there are extensive porto-systemic anastomoses. The presence of varices at other sites, such as the duodenum, small bowel and colon, the so called ectopic varices, are rare and usually follow sclerotherapy, oesophageal transection, paraoesophageal-gastric devascularization and extrahepatic obstruction of the portal vein. Ectopic varices due to portal hypertension are very rarely found in the small bowel and transverse colon. The hypothesis is that at these sites there is no porto-systemic communication, unless adhesions develop following a laparotomy. If they are present, they are due to increased venous pressure. Duodenal varices may occasionally coexist with oesophageal and gastric varices in patients suffering from cirrhosis. In this case they represent manifestations of porto-systemic communications in the retroperitoneum and kidney via pancreaticoduodenal and pyloric veins. However, if they are found in extrahepatic obstruction of the portal vein or SMV thrombosis, then they represent collateral circulation through the pancreatico-duodenal and pyloric veins. The most central of these para-duodenal veins enters the portal vein near its bifurcation.

Apart from SMV thrombosis and portal hypertension, ectopic varices can also be present throughout the GI tract in patients with angiodyplasia or haemangioma. They may be idiopathic or congenital in origin and secondary to arterio-venous fistula or pancreatitis.

As far as surgical management is concerned, this will depend on the patient’s clinical presentation. In cases of acute abdomen due to either intestinal obstruction or vascular occlusion, surgical intervention needs to be prompt, before ischaemic lesions become irreversible. Resection of the diseased bowel is the treatment of choice. If necessary, it is followed by the Ladd’s procedure, that is reduction of the twisted intestine in a counterclockwise direction and division of Ladd’s bands. The bands stretch across the second part of duodenum and the right abdominal wall, causing obstruction of the duodenum, as in our patient. In patients with chronic symptoms and in those where the pathology is an incidental finding, Ladd’s procedure is applied as well. These manoeuvres expose the base of the mesentery, release the caecum with the ascending colon and decompress the second part of the duodenum. Some authors advocate that it is not necessary to suture the caecum and the ascending colon in their new location. Others however recommend pexy if it is impossible to sufficiently broaden the base of the mesentery. If no pexy is performed then the procedure should be followed by appendicectomy to avoid the difficult problem of having a left-sided appendicitis. Laparoscopy, apart from being a useful tool in diagnosis, can also be the method of treatment since Ladd’s procedure can be performed laparoscopically.

Thrombosis of the SMV results in the formation of varices formation and bleeding from the GI tract. In these patients decompression of the SMV system should also be performed. A peripheral mesenteric branch of adequate caliber is anastomosed to the inferior vena cava, establishing a shunt that will prevent future episodes of bleeding. There is no need of anticoagulants.
This report describes an unusual cause of gastro-intestinal bleeding. Midgut volvulus, although a rare condition in adults, should be included in the differential diagnosis of ectopic varices with SMV thrombosis.

REFERENCES