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#### Case Report

# Major vascular and pancreatic penetrating trauma in patient with pre-existing MALS

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#### SUMMARY

We report a case regarding a 24-year-old male with severe intraabdominal complex vascular lesions involving the superior mesenteric vein (SMV), portal vein (PV), inferior vena cava (IVC), the Common Hepatic and Gastroduodenal arteries (CHA and GDA) in combination with a total transection of the pancreatic neck following a single penetrating trauma to the upper abdomen.

The management of the splanchnic vascular lesions were further complicated by the patient having pre-existing vascular condition known as Median Arcuate Ligament Syndrome (MALS). The MALS aspect makes this case unique and interesting to all trauma surgeons. Furthermore, this case report also reflects on the management of severe pancreatic trauma in the setting of the aforementioned vascular lesions.

### Case presentation

The patient was admitted to a level 1 Trauma Center in a circulatory stable condition. One major stab wound was seen in the upper abdomen with obvious communication to the peritoneal cavity. During laparotomy the liver lesion was observed but did not require any intervention. A contained non-expanding hematoma surrounding the pancreas retroperitoneum was present. It was decided not to explore the hematoma as the patient was circulatory stable. An 18Fr external drain was placed in the omental bursa.

An immediate postoperative CT scan revealed a total transection of the neck of the pancreas (Fig. 1) and an arterioportal (AP) fistula presumably arising from the CHA. The CT-scan also suspected injuries of the SMV/PV confluence and the IVC. Lastly the CT scan revealed a configuration of the celiac trunk which was consistent with MALS.

The patient remained circulatory stable. The drain production was with high levels of amylase. The AP fistula was attempted closed by insertion of an endovascular stentgraft (BeGraft  $6 \times 18$  mm, Bentley Innomed GmbH) in the CHA across the origin of the fistula. The arteriography revealed reverse flow through the GDA giving further evidence to the clinical importance of MALS in this patient (Fig. 2).

At POD 2 the patient was discussed with senior hepatobiliary surgeons. As the patient remained stable, it was decided to perform a relaparotomy with an intended resection of the pancreas. The risk of sudden and severe spontaneous hemorrhage was deemed very high due to the severe vascular lesions near the pancreatic transection with considerable leakage from the main pancreatic duct.

The first step during the relaparotomy was to access the IVC. This was done through a generous Kocher Maneuver exposing the IVC lesion which was subsequently sutured.

Intraoperative doppler ultrasound revealed a persistent AP fistula arising from the root of the GDA (Fig. 3). In order to close this fistula, the Proper Hepatic Artery (PHA) was clamped and divided above the GDA and the stentgraft was removed with 1 cm of the

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Fig. 1. CAT-scan after emergency laparotomy revealing a total transection of the pancreas (a). The CAT-scan also showed significant amount of blood in the abdomen. The complex vascular lesions are not presented in this image.

CHA. An anastomosis between the PHA and CHA was performed with 6-0 Prolene. Due to the pre-existing MALS condition the liver mainly received its arterial blood supply from the Superior mesenteric artery via the GDA. Division of the GDA thus resulted in a functional de-arterilisation of the liver. This was confirmed by doppler ultrasound. Due to the suspicion of MALS the CHA was dissected

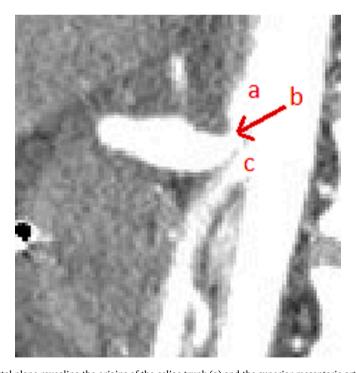


Fig. 2. CT-angiography in sagittal plane revealing the origins of the celiac trunk (a) and the superior mesenteric artery (c). The arrow "b" points to the significant deformation of the celiac trunk which is consistent with the median arcuate ligament syndrome.

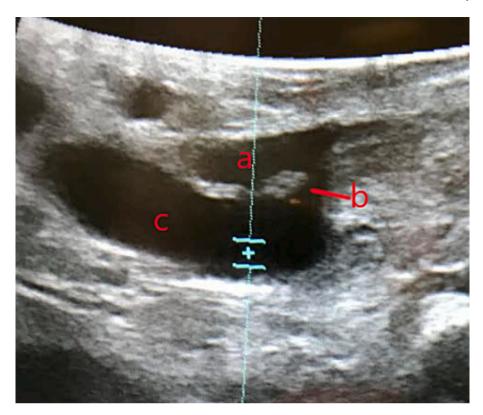


Fig. 3. Perioperative ultrasonography showing the gastroduodenal artery (a), the portal vein (c) and the traumatic arterioportal fistula between the two vessels (b).

back to the celiac trunk and further back to its origin from the aorta. Upon this dissection the median arcuate ligament and the diaphragmatic crura was divided freeing up the celiac trunk at its origin from the aorta. This dissection resulted in a sudden onset of adequate pulsatile flow in the celiac trunk and through the anastomosis between the CHA and PHA. A characteristic bi-phasic arterial doppler signal was observed in the liver.

Further exploration of the hepatoduodenal ligament and the transected pancreatic neck resulted in substantial bleeding from the PV/SMV confluence. The SMV, PV and splenic veins were clamped to gain access to the SMV/PV. This revealed a 3 cm long laceration in the anterior and posterior walls of the PV/SMV confluence with no possibility of simple suturing. The lacerated part of the PV/SMV was resected and an end-to end anastomosis of the PV and SMV was performed with 5-0 Prolene.

Due to the localization of the traumatic pancreatic transection and the presence of two vascular anastomoses a total pancreatectomy and splenectomy was carried out. The total operating time was 413 min and total perioperative blood loss 8500 ml.

The patient was hospitalized for 18 days after the relaparotomy and was afterwards transferred to the Department of Endocrinology with the primary focus on diabetes treatment adjustment.

#### Discussion

Lesions of the mesentericoportal veins are associated with a reported mortality ranging from 50 to 70 % [1]. And this mortality rate has been constant over the years [2]. The reasons for this are many but most obviously the SMV and PV are high flow veins which are likely to lead to exsanguination. In this case report the vascular lesions were constricted by the surrounding tissue forming a hematoma making the patient accessible for definitive surgery. Lesions of the PV and SMV are also located near other organs which is related to a considerable trauma-related morbidity such as the pancreas, duodenum, hepatoduodenal ligament and the liver.

The patient also presented with an AP fistula. According to Guzman et al. this fistula would be classified as a type B, thus requiring treatment [3]. An untreated grade B AP fistula carries risks both in the short and long term. In the short term there is a severe risk of bleeding and long term it would lead to portal hypertension and liver dysfunction and in some case obstruction of splanchnic venous flow through the liver causing bowel ischemia.

In this case it was decided to do a pancreatic resection due to the complex vascular lesions near the pancreatic lesion. A total pancreatectomy was chosen due to the extent of the vascular injuries and the resulting vascular reconstructions as a leakage from an eventual pancreatic anastomosis would carry a high risk of vascular complications such as bleeding/pseudoaneurisms, and thrombosis.

The patient had pre-existing MALS. In general, the very existence of MALS as a disease entity has been thoroughly debated [4]. The incidence of MALS is estimated as 2 per 100,000 patients [5]. The main hypothesis is that the symptoms are caused by decreased blood

flow through the celiac trunk due to extraluminal compression from the median arcuate ligament. This compression shows in a recognizable way upon imaging (CT-angiogram, MR-angiogram, Dupplex US). The treatment of MALS is to relieve the celiac trunk of it adherent connective tissue, thus improving the flow through the celiac trunk. There is a large group of patients with a radiological configuration of the celiac trunk which is consistent with MALS that has no symptoms (4). Thus, treatment of MALS is not indicated solely due to the radiological findings. In the cases of asymptomatic MALS the lack of symptoms is likely due to the collateral arterial flow through the gastro-and pancreatoduodenal arcades. This increased flow through collateral arteries has been hypothesized to cause secondary aneurisms which can lead to severe bleeding. In these cases, treatment of the aneurisms and the MALS is indicated [6,7]. The MALS condition in this case had to be managed operatively in order to re-establish arterial blood supply to the liver as the GDA, responsible for liver blood flow, had to be sacrificed due to the AP fistula. The physiology of this case was very much similar to Yamamoto et al. who conducted a pancreatoduodenectomy in the presence of MALS [8].

Conclusively this case report underlines the need for centralization to high-volume hepato-pancreato-biliary units with access to highly specialized HPB surgeons, anesthesiologists and interventional radiologists. To our knowledge there has never been published a case where the patient survived in the presence of a significant AP-fistula, severe SMV/PV lesion, lesion of the IVC and total transection of the pancreas in the presence of MALS.

#### CRediT authorship contribution statement

None.

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