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CASE REPORT

Ruptured Pancreaticoduodenal Artery Aneurysm with Median Arcuate Ligament Compression: A Two Staged Approach to Management

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Introduction: Pancreaticoduodenal artery (PDA) aneurysms represent a small portion of rare visceral artery aneurysms. Rupture of these aneurysms results in fatal haemorrhage in up to 50% of cases, necessitating prompt endovascular or open intervention. As highlighted by a recent retrospective review, median arcuate ligament (MAL) release is an important part of management when these aneurysms are diagnosed in conjunction with median arcuate ligament compression (MALC). Two cases of successful urgent management of a ruptured inferior pancreatoduodenal artery aneurysm with staged MAL release are reported.

Report: A 65 year old male presented with a ruptured PDA aneurysm in the context of MALC. The patient was treated by emergency transcatheter arterial embolisation (TAE). Staged laparoscopic MAL release required open conversion and stenting one month after rupture. A 73 year old male presented to the same institution with a ruptured PDA aneurysm, again in the context of MALC. This patient was similarly managed by emergency TAE and later had an uncomplicated laparoscopic MAL release. On table mesenteric angiography confirmed successful release. Both patients have since recovered without any recurrence of bleeding or new aneurysm formation.

Discussion: Ruptured true PDA aneurysms, while uncommon, may be managed successfully using urgent endovascular techniques. Concomitant coeliac axis stenosis due to MALC requires secondary treatment and can be managed effectively using a staged approach following the urgent presentation.

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INTRODUCTION

Pancreaticoduodenal artery (PDA) aneurysms represent a rare subtype of visceral arterial aneurysms. True PDA aneurysms arise from altered blood flow and pressure within pancreaticoduodenal arcades secondary to a proximal coeliac axis (CA) stenosis or occlusion.[1] Median arcuate ligament compression (MALC) is one cause of CA stenosis.[1,2] Treatment of PDA aneurysms at the time of diagnosis is recommended due to the high mortality rate following rupture.[2,3] Recent literature highlights the importance of MALC release for unruptured PDA aneurysms in an elective setting.[4] The management of two patients with ruptured inferior pancreaticoduodenal artery aneurysms and MALC using a two staged approach is presented.

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REPORT

Patient 1

A 65 year old male presented to the emergency department of a tertiary institution with acute onset epigastric pain. His history was significant for paroxysmal atrial fibrillation (AF) on apixaban, a pacemaker, and hypercholesterolaemia. On examination, he was haemodynamically stable with tenderness maximal over his epigastrium and right upper quadrant. His pathology demonstrated a haemoglobin level of 12.9 g/dL, with normal leucocyte count, liver function, and lipase. An abdominal computed tomography angiography (CTA) scan demonstrated an aneurysm in the anterior inferior pancreaticoduodenal artery (IPDA) measuring 9 mm craniocaudally with associated peri-pancreatic haemorrhage at the uncinate process.

Urgent endovascular treatment using a retrograde right femoral 6F (Brite-tip) sheath was performed. A diagnostic mesenteric digital subtraction angiogram (DSA) via a C2 catheter confirmed the presence of the aneurysm, but without active bleeding. Also noted was hypertrophy of the gastroduodenal artery, a CA stenosis, and reversal of flow through the pancreaticoduodenal arcade via the superior

mesenteric artery (SMA) (Fig. 1A). Selective anterior IPDA coil embolisation from the SMA approach was performed via a 2.4 French (Progreat-Terumo) microcatheter within a C2 catheter using two 3 × 15 mm ruby coils (Penumbra). Despite some coil prolapse into the origin of the posterior IPDA, there was no residual filling of the aneurysm and all other arcades remained patent (Fig. 1B).

Post-operatively, an autoimmune screen, vasculitis studies, and infectious serology were normal. At one month, a staged laparoscopic MAL release was performed to allow time for the retroperitoneal haematoma to resolve. Despite adequate laparoscopic visibility and multidisciplinary consensus of visual release, post-operative CTA showed a residual 75% CA stenosis (Fig. 1C). Coeliac trunk stenting was initially attempted, but due to poor stent expansion from the residual MAL fibre an open release was necessary. Once the remaining band was released the stent was re-angioplastied (Fig. 1D). Anticoagulation was restarted on discharge at day 7. Follow up imaging at 12 months revealed a widely patent CA stent and no further PDA aneurysm formation.

Patient 2

A 73 year old male presented with a two hour history of acute upper abdominal pain on a background of AF with apixaban use. His history was significant for previous coronary stenting and dyslipidaemia. On examination, the patient was hypotensive to 97/60 mmHg and had a pulse rate of 75 beats/min. He had mild tenderness in the epigastrium without peritonism. His pathology demonstrated a haemoglobin level of 9.8 g/dL and normal leucocyte count, liver function, and lipase. Abdominal CTA identified a ruptured aneurysm arising from a branch of the anterior IPDA close to its origin, as well as a proximal CA stenosis (Fig. 2C).

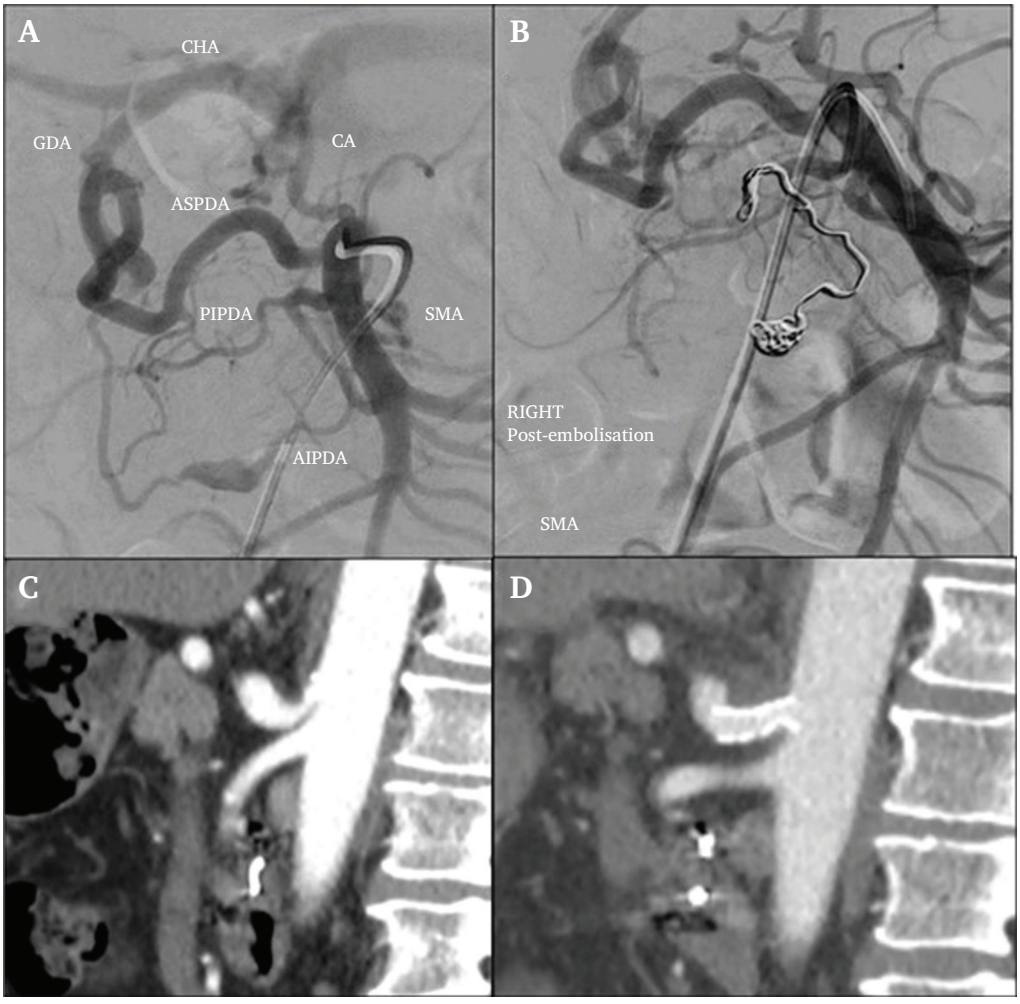

Figure 1. (A) Patient 1. Anterior inferior pancreaticoduodenal artery (AIPDA) aneurysm with hypertrophy of gastroduodenal artery (GDA) and reversal of flow through the pancreaticoduodenal arcade via the superior mesenteric artery (SMA) suggesting proximal coeliac trunk stenosis marked CA (coeliac axis). Common hepatic artery (CHA), posterior inferior pancreaticoduodenal artery (PIPDA), and anterior superior pancreaticoduodenal artery (ASPDA) marked for reference. (B) Patient 1. Post successful coil embolisation of the inferior pancreaticoduodenal artery (IPDA) aneurysm. (C) Patient 1. Computed tomography angiography sagittal section showing proximal coeliac stenosis with retroperitoneal haemorrhage associated with IPDA aneurysm rupture, prior to median arcuate ligament compression release. (D) Patient 1. Final sagittal plane image of patient 1 CA post-release and stenting of the residual coeliac stenosis.

Urgent coil embolisation of the bleeding branch of the IPDA was performed, accessed by a 6 French sheath in the right femoral artery (Fig. 2A). A C2 catheter was used for SMA cannulation, and this platform allowed five 2 × 4 cm ruby coils to be deployed via the 2.4F (Progreat-Terumo) microcatheter (Fig. 2B). The whole IPDA remained patent following selective branch coiling. The patient had an uncomplicated recovery and was discharged on day 6.

Three months after discharge, laparoscopic MAL release in a hybrid operating theatre with completion mesenteric angiography was undertaken. A second preparation and draping was required following laparoscopic port closure under the same anaesthetic. Owing to the experience with the first patient, intra-operative assessment of release was performed simultaneously to facilitate immediate open conversion if required. An on table mesenteric angiogram via a right femoral 6 French sheath and C2 catheter demonstrated restored antegrade CA flow with less than 30% residual CA stenosis. Therefore, open surgery was not required. At the patient's request, a post-operative mesenteric duplex ultrasound was performed instead of CTA, which confirmed antegrade flow and a widely patent CA at three months (Fig. 2D).

DISCUSSION

PDA aneurysms characterise 2% of reported visceral arterial aneurysms, with diagnosis occurring at a mean age of 60 years.[1,5] Life threatening haemorrhage can occur where these aneurysms rupture.[5,6] Rupture presents with broad clinical signs, ranging from vague abdominal pain and nausea to syncope and haemorrhagic shock.[5,6] Abdominal CTA is the most common modality for PDA aneurysm

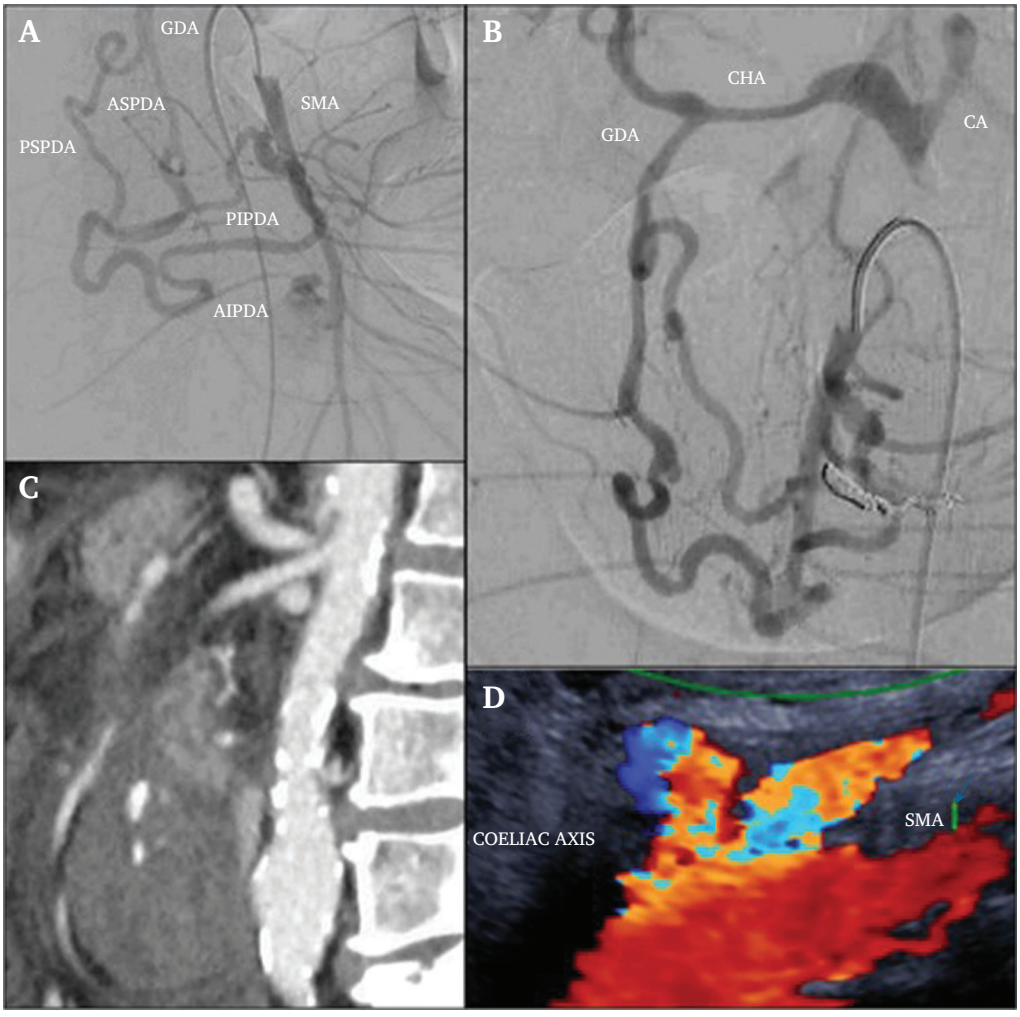

Figure 2. (A) Patient 2. Digital subtraction angiography showing the bleeding inferior pancreaticoduodenal artery (IPDA) aneurysm. (B) Patient 2. Post-endovascular coiling with retrograde filling of the pancreaticoduodenal arcade and proximal coeliac axis (CA). Common hepatic artery (CHA) and gastroduodenal artery (GDA) have been marked for reference. (C) Patient 2. Computed tomography angiography sagittal scan prior to treatment showing proximal coeliac stenosis with large retroperitoneal haemorrhage associated with IPDA aneurysm rupture. (D) Patient 2. Post-intervention duplex ultrasound showing antegrade flow in CA and superior mesenteric artery (SMA). AIPDA = anterior inferior pancreaticoduodenal artery; ASPDA = anterior superior pancreaticoduodenal artery; PIPDA = posterior inferior pancreaticoduodenal artery; PSPDA = posterior superior pancreaticoduodenal artery.

diagnosis.[5] The goals of treatment are not only haemorrhage control, but also to identify and treat any underlying pathology contributing to aneurysm formation.[2,3,6] In these cases, the secondary aim is to re-establish antegrade CA flow to depressurise the pancreaticoduodenal arcade.

The presence of CA stenosis has been observed in 50%—80% of patients with true PDA aneurysms.[3—6] Sutton and Lawton (1973)[2] first proposed that CA stenosis increases retrograde flow in the peri-pancreatic arterial networks via the SMA, leading to the formation of true PDA aneurysms.[2,5] Recent dynamic imaging studies have confirmed this effect.[4,6,7] Nishiyama et al. suggested that the inferior branch is more susceptible to aneurysm development when there is absent communication between superior and inferior branches in the PDA network.[3] Revascularisation of the CA allows for restoration of antegrade CA flow which can stabilise PDA aneurysms. Salomon du Mont et al. demonstrated ruptured PDA aneurysm resolution with CA bypass alone.[3,8] Given that multiple reports demonstrate that PDA aneurysm size does not predict rupture, active management of both the aneurysm and the CA stenosis is required for definitive treatment.[1,3—5,8,9]

The first stage of management should focus on the PDA aneurysm. Treatment approaches for ruptured PDA aneurysms with MALC vary from transcatheter arterial embolisation (TAE) with or without coeliac artery stenting to open aneurysmorrhaphy with or without concomitant MAL release.[1—4,9] Percutaneous TAE is considered the preferred first line option for both ruptured and unruptured PDA aneurysms.[2,4,6] TAE also allows treatment of the concomitant CA stenosis using stents. The risks of TAE include failure to access the PDA aneurysm and distal embolisation due to coil migration.[3,4,6] Hepatic or foregut ischaemia may occur after TAE where foregut perfusion is reliant on the coiled collaterals.[2,8,9] If urgent open surgery is required, reported techniques include PDA ligation, resection with bypass, or pancreaticoduodenectomy. In addition, open MALC release can be performed simultaneously.[3—5]

The second stage of treatment should address the MALC. The treatment of a causative coeliac stenosis due to MALC can be performed using an open or laparoscopic approach.[2,3,7,9] In patients with vessel fibrosis or CA occlusion, further treatment with a CA bypass or a CA stent may be required.[4,8] A recent retrospective study has cautioned the use of CA stenting alone for MALC treatment, with re-stenosis or occlusion occurring in up to 50% of cases in the short to medium term.[4] Illuminati et al. reviewed 57 patients with an unruptured PDA aneurysm with MALC. There was no PDA recurrence or recanalisation in those managed by open MALC treatment. Recanalisation however did occur in 11% following endovascular treatment and was correlated with CA re-stenosis.[4]

Stenting should be reserved for intrinsic causes of CA stenosis (atherosclerosis) or following extrinsic MAL release when antegrade flow is not achieved in the arcade. Antegrade flow can be confirmed via either palpation, intra-operative ultrasound, intra-operative angiography, or on post-operative duplex.[5,7] The first of the patients had noticeable improvement in the palpable pulse at open release, while the second demonstrated restoration of antegrade flow on DSA compared with the emergency presentation images.

Staged MALC release was performed one to three months following the PDA aneurysm rupture to facilitate resolution of the retroperitoneal haematoma, which could impair operative MAL techniques.[3,8] Currently, there is no consensus on a preferred timeline for staged intervention and it remains dependent on patient factors and local access to treatment.[2,4,9] A report by Terayama et al. in 2019 advocated a similar approach with an extrinsic MAL release one month post-emergency TAE for a ruptured PDA aneurysm.[9]

CONCLUSION

True PDA aneurysms are rare, frequently fatal if ruptured, and can be associated with MALC as a cause of CA stenosis. In this case report, the two patients with ruptured PDA aneurysm and MALC were successfully managed with TAE as first line treatment to arrest haemorrhage, followed by a staged extrinsic release of the MALC. Treatment of underlying MALC is required for the effective correction of altered haemodynamics in the PDA arcade. When adequate collateral circulation allows, a staged MALC release offers a period of recovery from acute rupture and can improve operative success.

CONFLICT OF INTEREST

None.

FUNDING

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