

CASE REPORT

Vascular Plug Embolization of a Superior Mesenteric Arteriovenous Fistula: Case Report and Literature Review of Endovascular Treatment

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Introduction: Arteriovenous fistula (AVF) rarely occurs in the portal venous system. Aetiologies include iatrogenic, surgical, and penetrating trauma of the abdomen. Clinical manifestations of superior mesenteric portal arteriovenous fistula (SMPAVF) are right heart failure, mesenteric ischaemia, or signs of portal hypertension.

Report: The case of a 42 year old man with a history of Crohn's disease who had a delayed symptomatic mesenteric portal AVF, occurring 20 years after ileocecal resection, which was subsequently managed by endovascular approach is reported. The patient presented with post-prandial abdominal pain for almost one year, and dyspnoea New York Heart Association stage II. There were no signs of portal hypertension. Pre-operative contrast enhanced computed tomography showed a high flow SMPAVF, with a short and wide neck (9 mm × 16 mm) at the level of the last collateral of the superior mesenteric artery. It was associated with a large aneurysm of the mesenteric vein. Vascular plug embolisation (Amplatzer 18 × 18 mm, Abbott, Chicago, IL, USA) by femoral access allowed exclusion of the SMPAVF and preserved arterial flow in the distal collaterals. During follow up, the patient developed portal vein thrombosis and required therapeutic anticoagulation for six months.

Discussion: In most cases, endovascular approaches are preferred in the management of SMPAVF. Endovascular approaches are based on minimally invasive techniques including embolisation (coiling or plug) and covered stenting. Vascular plug embolisation of SMPAVF is feasible and seems to be an effective technique, with the advantage of saving collaterals. Therapeutic anticoagulation should be considered post-operatively in cases with venous dilatation and reduced flow linked to exclusion of the AVF.

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INTRODUCTION

Arteriovenous fistula (AVF) rarely occurs in the portal venous system. Localisations are hepatic portal AVF, splenic portal AVF, and rarely superior mesenteric portal AVF (SMPAVF).¹ Aetiologies include iatrogenic and penetrating trauma of the abdomen. SMPAVF may occur years after trauma or surgery and remains asymptomatic for a long time.² The main clinical manifestations of SMPAVF are right heart failure, mesenteric angina, or signs of portal hypertension.³ Due to the rarity of SMPAVF, there is no therapeutic consensus in the literature. However, endovascular treatment seems to be the preferred management in the first instance. The objective of endovascular treatment is to exclude the fistula using a minimally invasive technique

such as embolisation or covered stenting. Embolisation can be performed with a coil or plug and requires catheterisation of the fistula neck. Covered stenting may cover collaterals and require antiplatelet therapy.

The main objective was to report the case of a patient with a delayed superior mesenteric portal AVF, focusing on an endovascular approach using vascular plug embolisation, and to review the literature on endovascular treatment.

CASE REPORT

A 42 year old man with a history of Crohn's disease presented with post-prandial abdominal pain lasting almost one year, suggestive of mesenteric angina. He presented with abdominal pain, first in the epigastric area and then in the left hypochondrium associated with diarrhoea. He described an increase in pain 10 minutes after a meal. In addition, the patient presented with dyspnoea (New York Heart Association stage II) in the prone position, which triggered cardiological exploration. Electrocardiogram, cardiac biological markers (N-terminal pro-brain natriuretic peptide = 61, norm <81.5 ng/L), and echocardiography (left ventricular ejection

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fraction: 60%) were normal. Contrast enhanced computed tomography (CT) showed a high flow SMPAVF, with a wide neck at the level of the last collateral of the superior mesenteric artery, and a large aneurysm of the mesenteric vein (maximum diameter = 66 mm). There were no signs of acute complications of Crohn's disease (Fig. 1).

The SMPAVF was probably related to the ileocecal surgery performed 20 years previously. Chronic inflammation due to Crohn's disease may also have been a contributory factor.

An endovascular approach was preferred due to the high morbidity and mortality risk with the open repair of such a large, complex fistula in a setting of Crohn's disease. Covered stenting was not performed because of the distal location of the AVF and the presence of multiple collaterals. The neck of the AVF was short and wide, increasing the risk of coil migration. Therefore, embolisation with a vascular plug was performed.

Through a right percutaneous femoral access, the superior mesenteric artery was catheterised with a guiding sheath (Cordis, JR4 6F 100, France) and a 0.035 guidewire. Intra-operative angiography revealed a high flow SMPAVF with a short and distal neck. Digital subtraction angiography (DSA) allowed precise measurements of the artery, vein, and fistula to be obtained. The fistula was short and wide (9 mm × 16 mm). The AVF was catheterised using a micro-catheter system (Progreat Micro Catheter System 2.4 Fr, Terumo, Tokyo, Japan). The AVF was catheterised with 5Fr catheters. (Impress Diagnostic Peripheral Catheters 5Fr, Merit, Salt Lake City, UT, USA). Vascular plug oversizing was 20%, which is the usual management for arterial and venous plugs. A vascular plug (Amplatzer 18 × 18 mm, Abbott, Chicago, IL, USA) was deployed at the level of the neck of the AVF between the mesenteric vein and the mesenteric artery, allowing exclusion of the AVF and restoration of arterial flow in the distal collaterals (Fig. 2).

Twenty-four hours post-operatively, the patient presented an episode of abdominal pain with diarrhoea and rectorrhagia, but no signs of hepatic failure. Laboratory findings were elevated, C reactive protein 36 mg/L (normal

range [N] < 5 mg/L), polymorphonuclear neutrophils 4.31 G/L (Giga/liter) (N = 1.7–7.5), haemoglobin 13.5 g/dL (N = 13–17.5), hepatic transaminase aspartate aminotransferase 29 IU/L (N = 10–50), and alanine aminotransferase 41 IU/L (N = 10–50). D dimer was not measured.

Post-operative CT showed complete exclusion of the SMPAVF. The superior mesenteric artery was patent, with no signs of mesenteric ischaemia. However, thrombosis of the mesenteric vein was observed, extending to the portal vein (Supplementary Video S1). Therapeutic anticoagulation by low molecular weight heparin (enoxaparin 200 IU/kg/24 hours in two subcutaneous injections) was administered during hospitalisation. On day 11, the patient was discharged and therapeutic anticoagulation was switched to warfarin 4 mg daily.

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The following is the supplementary data related to this article: Video S22

At three months, the patient had no abdominal pain and no rectorrhagia or other symptoms. Laboratory findings were normal. Follow up CT showed complete exclusion of the SMPAVF. A reduction in size of the endoluminal defect (47 × 40 mm vs. 53 × 45 mm) and partial patency of the portal vein were observed (Supplementary Video S2). Due to persistence of thrombosis, therapeutic anticoagulation was maintained with warfarin four mg daily (Fig. 3).

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.ejvsf.2023.08.001>.

The following is the supplementary data related to this article: Video S33

DISCUSSION

SMPAVF are extremely rare; there have been fewer than 50 reports in the literature. A descriptive analysis of the studies retrieved in the literature review is presented in Table 1. Clinical manifestations, causes, radiological diagnosis, intervention, and outcomes are summarised in Supplementary Table S1. SMPAVF are most often the result of traumatic or iatrogenic injury. Diagnosis is usually delayed because SMPAVF is often asymptomatic for many years. An early diagnosis is more frequent in cases of traumatic injury. The most frequently described clinical symptom is mesenteric angina with abdominal pain and diarrhoea following meals. Less commonly, increased portal venous pressure gives rise to hepatic congestion, venous aneurysm, ascites, and oesophageal varices.⁴ The development of oesophageal varices leads to bleeding.⁵ The next stage of the disease is right heart failure. In fact, delayed presentation is an independent predictive factor of right heart failure.⁶ This justifies a cardiological assessment before treatment, as was done in this case.

Imaging is a useful tool for diagnosis, characterising the SMPAVF and guiding management. DSA is the gold standard examination to show the exact anatomical location, fistula flow, and dynamics as well as analysis of collateral branches. However, this procedure is typically used only for the



Figure 1. Pre-operative computed tomography. ★ = superior mesenteric vein; ☆ = superior mesenteric artery; ▽ = fistula.

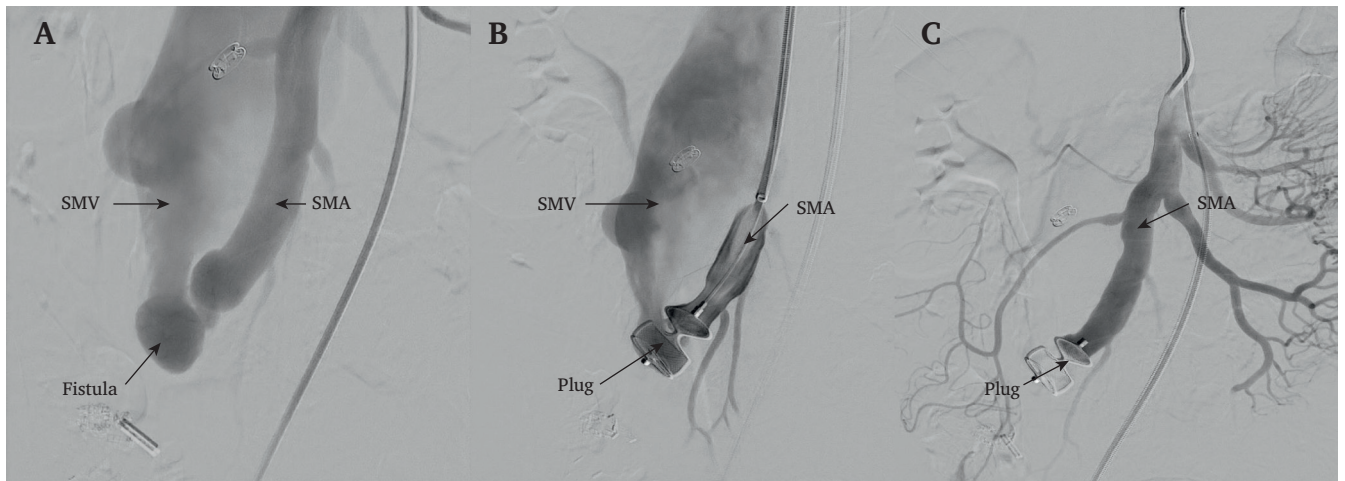


Figure 2. Intra-operative and post-operative angiography. SMV = superior mesenteric vein; SMA = superior mesenteric artery. (A) Superior mesenteric arteriovenous fistula (SMAVF) with SMV aneurysm. (B) Vascular plug (Amplatzer 18 × 18mm) deployment. (C) Post-operative angiography confirmed complete exclusion of the arteriovenous fistula (AVF).

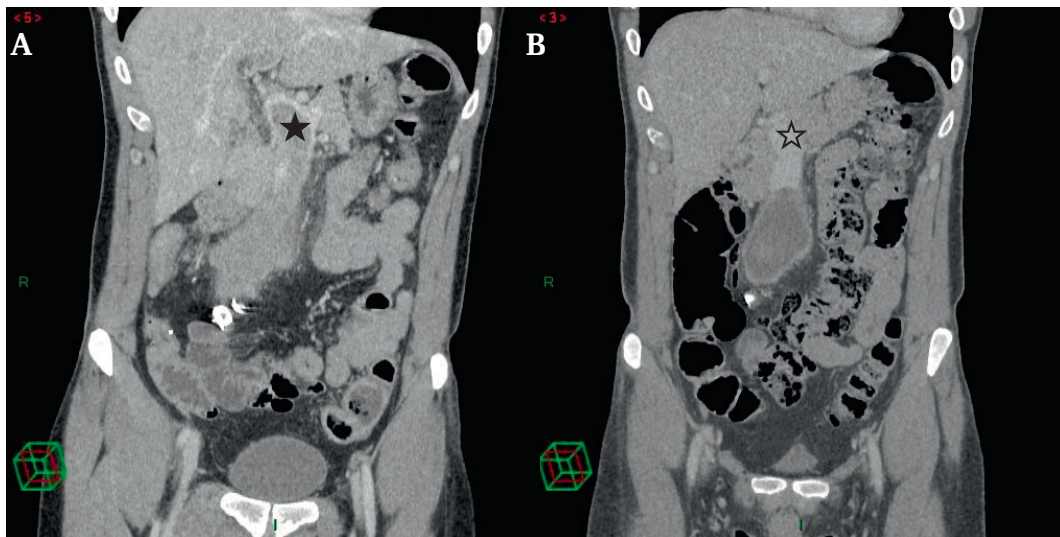


Figure 3. Post-operative computed tomography. (A) Thrombosis of the superior mesenteric vein, extending to the portal vein (★). (B) At three months, reduction in size of the thrombus and limited to the superior mesenteric vein. Patency of the portal vein (☆).

treatment. CT contributes to diagnosis and to treatment strategies. In some cases, Doppler ultrasound can be a useful tool to assess the haemodynamic impact but is difficult and requires an experienced operator.

In this case, the patient presented typical symptoms: mesenteric angina, portal congestion with portal system aneurysm, and signs of right heart congestion with dyspnoea. CT scan can show the anatomical location of the SMPAVF, which in this case was distal with a short and wide neck.

Before the 2000s, the majority of cases of SMPAVF were treated by open repair surgery, associated with laparotomy and general anaesthesia, with good surgical outcomes. Today, endovascular approaches, based on minimally invasive techniques, are preferred in the management of SMPAVF, as

they seem to be associated with lower morbidity and mortality, even though no study has compared them.

The most commonly used embolisation technique is coil embolisation (20 cases), but there is a risk of coil migration and distal mesenteric arterial occlusion, or portomesenteric venous thrombosis. This risk may decrease if the fistula neck is long and narrow, and the flow rate is not too high.

Many cases have been managed successfully using covered stents (nine cases). Covered stenting represents an effective and reproducible technique. This technique seems more used when the anatomical location is proximal (90% cases). However, endovascular stent placement may cover collateral arteries⁷ and carries an increased risk of thrombosis, requiring long term antiplatelet therapy.⁸

Table 1. Descriptive analysis of the studies retrieved in the literature review

Characteristics	
Mean age – y	41 ± 15
Male sex	33 (71.8)
Aetiology	
Surgery history	30 (65.3)
Trauma history	16 (34.7)
Symptoms	
Mesenteric angina	45 (97.8)
Right heart failure	7 (15.2)
Portal hypertension	25 (54.3)
Diagnostic imaging	
DSA	27 (58.7)
US	10 (21.7)
CT	33 (71.7)
Localisation	
Proximal	21 (45.7)
Distal	23 (50)
Unspecified	2 (4.3)
Intervention*	
Open repair	14 (30.4)
Stent	9 (19.6)
Coils	20 (43.4)
Plug	4 (8.8)
Outcomes	
Success	
Open repair	13 (92.8)
Stent	9 (100)
Coils	19 (95)
Plug	4 (100)
Death	
Open repair	1 (7.2)
Stent	0
Coils	1 (5)
Plug	0
Portal thrombosis	
Open repair	0
Stent	0
Coils	5 (25)
Plug	1 (25)

Data are presented as *n* (%) or mean ± standard deviation. DSA = digital subtraction angiography; US = ultrasound; CT = computed tomography.

* One patient died before treatment, one patient was treated by stenting and coils and one patient was treated by coils and plug.

Vascular plug embolisation is rarely used (four cases), but seems to be an effective technique, particularly when the neck of the fistula is wide. The main advantage of using vascular plug embolisation, rather than stenting, is that it can save some collaterals and it does not require antiplatelet therapy.

An endovascular first approach was used because of the patient's history of Crohn's disease and laparotomy. Imaging findings and the anatomy of the SMPAVF guided the choice of technique. A covered stent could not be used because of the distal anatomical location of the SMPAVF. Potential migration of the vascular plug was less of a worry because 20% oversizing was used. The use of a coil might have increased the risk of migration and losing the catheterisation.

In this case, the main advantages of using a plug over other embolic devices were better release accuracy and a lower migration risk.

Venous thrombosis is a potential complication of venous dilatation and reduced flow linked to the exclusion of AVF.⁹ There is no consensus in the literature on the prevention of thrombosis after exclusion of AVF, but six cases of portal vein thrombosis after SMPAVF exclusion have been described. The risk of thrombosis seems increased in the presence of a venous aneurysm and can be managed post-operatively with therapeutic anticoagulation. It is possible that therapeutic anticoagulation should have been considered at the time of the procedure because the patient had no contraindication and there was a large aneurysm of the mesenteric vein. In this case, anticoagulant treatment was able to wash out the portal thrombosis.¹⁰

Conclusion

SMPAVF is extremely rare and diagnosis is complicated because, in most cases, patients remain asymptomatic for a long time. Endovascular approaches, based on minimally invasive techniques such as vascular plug embolisation or covered stenting, can be used to treat SMPAVF according to anatomical location, with good outcomes.

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CONFLICT OF INTEREST

None.

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APPENDIX A. SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.ejvsf.2023.08.001>.

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