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CASE REPORT

A Case of Refractory Esophageal Varices Caused by an Inferior Mesenteric Arteriovenous Malformation with All Portal System Occlusion Successfully Treated via Transarterial Embolization

Natsuhiko Saito[1)], Masayoshi Inoue[1)], Kentaro Ishida[1)], Hidehiko Taguchi[1)], Masayo Haga[1)], Emiko Shimoda[1)], Kengo Morimoto[1)], Junko Takahama[1)] and Toshihiro Tanaka[2)]

1) Department of Radiology, Higashiosaka City Medical Center, Japan
2) Department of Diagnostic and Interventional Radiology, Nara Medical University, Japan

Abstract:
Inferior mesenteric arteriovenous fistulas/malformations are rare, reported in only 40 cases as of 2021. Their main manifestations include portal hypertension and ischemic bowel disease. We report the case of a 50-year-old man with refractory esophageal varices caused by this condition that was successfully treated with transarterial embolization. Computed tomography revealed an inferior mesenteric arteriovenous malformation and ascending blood flow into the esophageal varices through a remarkably dilated marginal vein. All portal systems were occluded, possibly because of the myointimal hyperplasia of the inferior mesenteric vein. The patient recovered without hemorrhagic events after transarterial embolization and endoscopic injection sclerotherapy. This is the first report of an inferior mesenteric arteriovenous malformation resulting in refractory esophageal varices with all-portal system occlusion successfully treated with transarterial embolization.

Keywords:
arteriovenous mesenteric fistula, arteriovenous mesenteric malformation, portal system occlusion, transarterial embolization, esophageal varices

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Introduction

Inferior mesenteric arteriovenous fistulas or malformations (AVFs/AVMs) are rare and can be congenital, idiopathic, traumatic, or iatrogenic in etiology [1, 2]. Iatrogenic AVFs can occur secondary to penetrating abdominal arterial catheterization or surgery, such as sigmoidectomy or left hemicolectomy [1]. Conversely, congenital or idiopathic etiologies are associated with the Osler-Weber-Rendu syndrome, the Ehlers-Danlos syndrome, or an unknown etiology. Congenital AVFs can originate from embryonic vessels that fail to regress or from collagen defects [1]. As of 2021, only 40 cases, involving 25 primary and 15 secondary cases, have been reported [2]. The main manifestations of this condition include portal hypertension and ischemic bowel disease. Symptoms include abdominal pain, weight loss, diarrhea, tenesmus, nausea, vomiting, thrill and mass, lower and upper gastrointestinal bleeding, ischemic colitis, and heart failure [1, 5-7]. Although treatment strategies involve surgical intervention or embolization [3], there are few reports of symptomatic recovery with transarterial embolization (TAE). Inferior mesenteric AVFs/AVMs are often associated with inferior mesenteric vein (IMV) occlusion [1], but cases of their association with all-portal system occlusions have not been reported. In this report, we present the first case of refractory esophageal varices, possibly caused by an inferior mesenteric AVM with all-portal system occlusion, which was successfully treated with TAE.

Case Report

The patient was informed of the details of this case report, and written informed consent for the use of medical records (including the use of images) was obtained. Institutional Review Board approval was not required for this case report according to the ethical guidelines of our institution,

Corresponding author: Natsuhiko Saito, summernatsu@naramed-u.ac.jp
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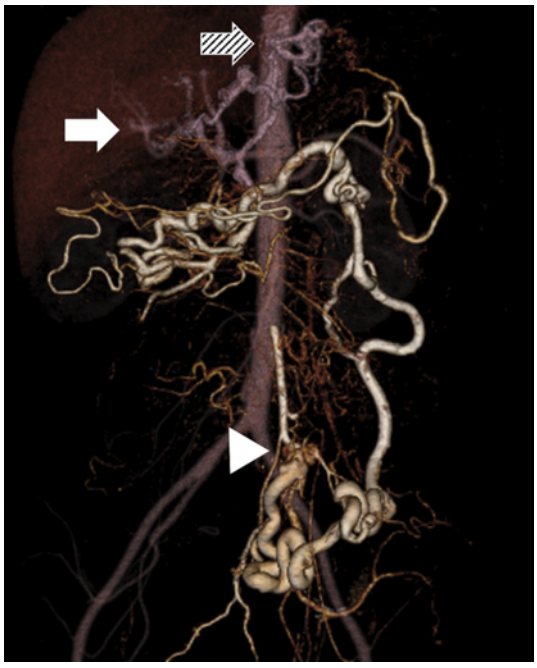

Figure 1. Three-dimensional contrast-enhanced computed tomography revealed an inferior mesenteric AVM (arrowhead) and ascending blood flow into the slightly patent superior mesenteric vein (SMV) divided into esophageal varices (diagonal arrow) and the fine collateral vessels to the liver (white arrow) through a remarkably dilated marginal vein. All portal systems, including the portal vein, most segments of the SMV, splenic vein, and IMV, were occluded.

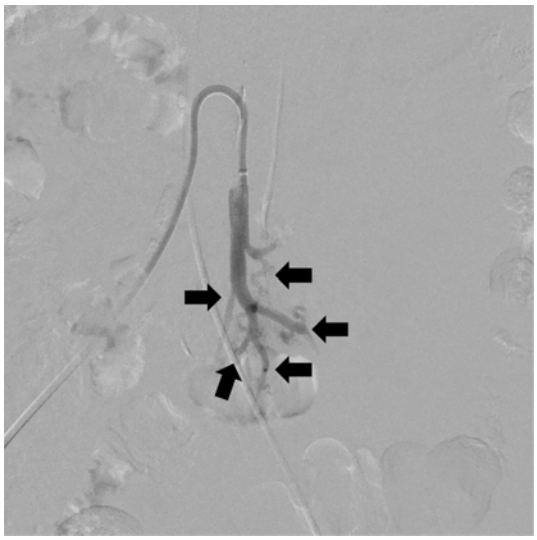

Figure 2. Digital subtraction angiography (DSA) from the IMA. Five feeding vessels arising from the IMA into the IMV are shown (arrows). On the later phase, these five feeding vessels further branched into countless fine vessels like nidus into the IMV (not posted).

and written informed consent related to TAE treatment was obtained from the patient before the procedure.

A 50-year-old man was referred to our gastroenterology department for treatment of esophageal varices. The patient had no history of abdominal surgery or trauma. The patient's chief complaint was abdominal distension. He had experienced melena several times before arriving at our hospital. He was a drinker and had neither hepatitis B nor C virus infection. Previous noncontrast-enhanced computed tomography (CT) showed hepatic atrophy and splenomegaly, and blood test revealed severe anemia. Upper gastrointestinal endoscopy performed at our institution showed worsening of the esophageal varices (LsF2CwRC3). Endoscopic injection sclerotherapy (EIS) was attempted to treat the varices, but EIS failed because of fast blood flow in the varices. Therefore, endoscopic variceal ligation was performed to prevent hematemesis, but hematemesis occurred 3 days later. Contrast-enhanced CT, performed to identify the source of bleeding, revealed an inferior mesenteric AVM and ascending blood flow into the slightly patent superior mesenteric vein (SMV) divided into esophageal varices and fine collateral vessels to the liver through a remarkably dilated marginal vein. All portal systems, including the portal vein, most segments of the SMV, splenic vein, and IMV, were occluded (**Fig. 1**). The patent part of the SMV was from the middle of L2 to the upper edge of L1 (～4-cm-long patency). Diagnostic angiography was performed for further investigation. Selective angiography revealed a vascular malformation, including five feeding vessels arising from the inferior mesenteric artery (IMA) into the IMV (**Fig. 2**), followed by a remarkably dilated marginal vein, which was connected to the esophageal varices (**Fig. 3a-3d**) and fine collateral vessels in the liver. These five feeding vessels further branched into numerous fine vessels in a nidus-like formation in the IMV. As observed in CT, the portal system was occluded and hepatic blood inflow could be secured by fine collateral pathways. Angiography from the splenic artery revealed that the blood from the spleen went through the short gastric vein to the esophageal vein. On the basis of these findings, an inferior mesenteric AVM with all-portal system occlusion was diagnosed. The following day, transarterial shunt embolization was performed. The right femoral artery was punctured, and a 4.5-Fr guiding sheath (Parent Plus; Medikit Japan, Tokyo, Japan) was placed in the IMA. To strengthen the backup, a 4-Fr guiding catheter (Cerulean G; Medikit Japan, Tokyo, Japan) was used in the guiding sheath and was placed near the shunts. For the selective catheterization of the five inflow vessels, a microcatheter with two markers (Excelsior 1018; Stryker Japan, Tokyo, Japan) and a 0.016" microwire (SUCCEDO; Boston Scientific Japan, Tokyo, Japan) were used. All five vessels forming the AVM were embolized using metallic coils (Target coils; Stryker Japan, Tokyo, Japan, C-STOPPER coils; PIOLAX Medical Device, Kanagawa, Japan, and Nester coils; Cook Medical Japan, G.K., Tokyo, Japan). Postembolization angiography revealed a drastic decrease in shunt flow, and intestinal blood flow was preserved. Postembolization upper gastrointestinal endoscopy revealed improvement in the esophageal varices, which were F1 with a drastic reduction in the red color sign (**Fig. 4a and 4b**). The shunt vessels were completely occluded on angiography performed 12 days after the TAE (**Fig. 5**). Because the inflow to the remaining

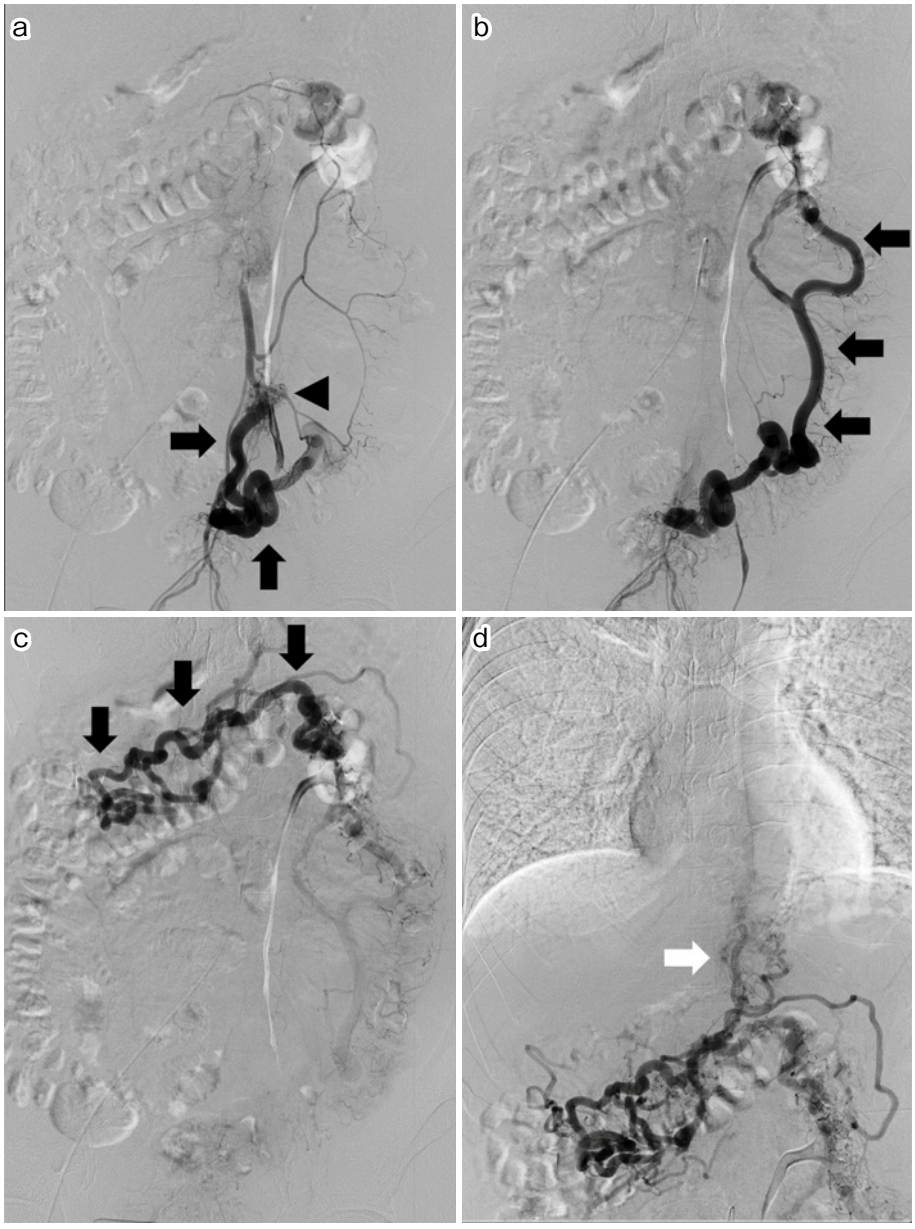

Figure 3. Digital subtraction angiography (DSA) from the IMA. The AVM (arrowhead) is a continuous remarkably dilated marginal vein (black arrows) that is connected to esophageal varices (white arrow). The phase has progressed from Figure 3a to Figure 3d.

esophageal varices was provided by the vessels from the short gastric vein to the esophageal vein via the gastric vein, EIS was performed under the balloon occlusion of the splenic artery to promote sclerosing agent stasis. The varices were successfully treated with good stagnation of a sclerosing agent. Liver function did not change after embolization. Child-Pugh score did not change from 9 to 9, reflecting a low blood albumin level (2.4→2.5) and a moderate amount of ascites. After the procedure, the patient was discharged at convenience and refused any additional treatment against the recommendation of partial splenic embolization. Fortunately, the patient was alive and returned to work ～4 months after self-discharge, with no hemorrhagic events.

Discussion

Inferior mesenteric AVFs/AVMs are rare. Until 2021, only 40 cases had been reported [3, 4]. Kai et al. stated that AVFs could be easily confused with AVMs because there are no criteria for distinguishing the former from the latter [4]. In the current case, we defined a diagnosis as an AVM when the vessels directly connected to the IMA and IMV were numerous and fine nidus-like. Although the main manifestations of this condition include portal hypertension and ischemic bowel disease, only two cases have revealed severe portal hypertension, contributing to the development of esophageal varices [5, 8]. Most patients present with ischemic bowel disease manifesting as abdominal pain and lower gastrointestinal bleeding [3]. Ischemic bowel disease is caused by venous stasis due to shunts and decreased arte-

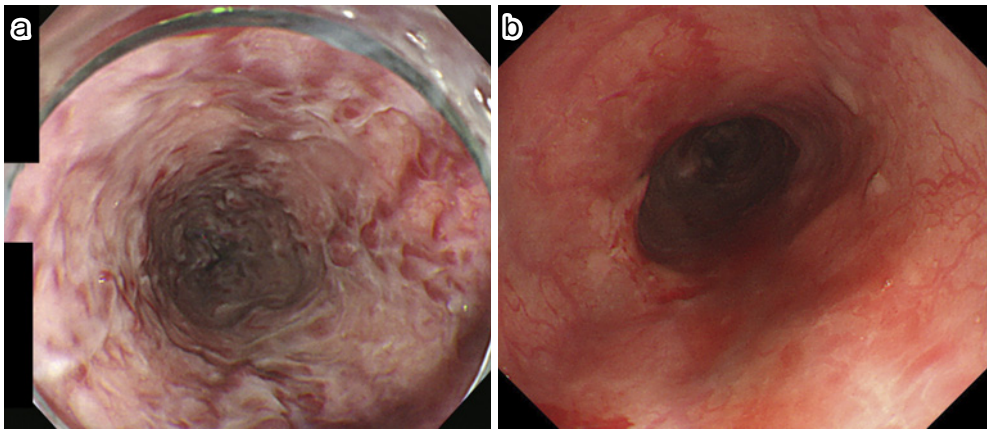

Figure 4.
a) An endoscopic image before treatment. Developed esophageal varices with much red color sign were observed.
b) An endoscopic image after treatment. Improvement of the esophageal varices, which are F1 with a drastic reduction in the red color sign, was observed.

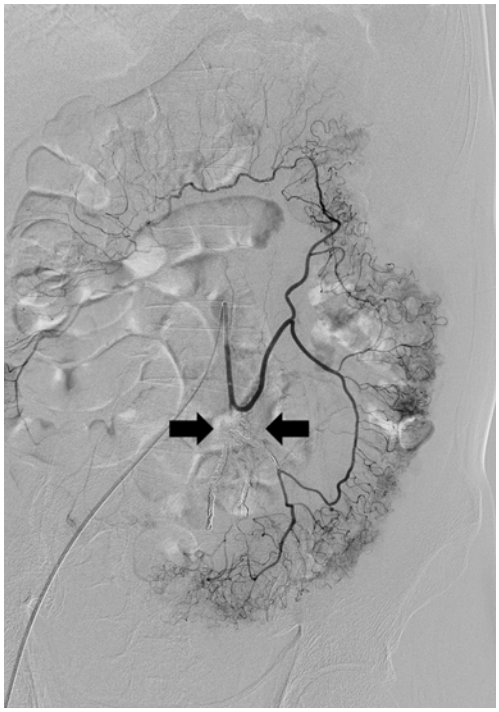

Figure 5. Digital subtraction angiography (DSA) from the IMA postembolization. Shunt vessels were angiographically completely occluded (arrows). The blood flow to the remarkably dilated marginal vein also disappeared. The intestinal blood flow was preserved.

rial flow to the bowel wall, resulting in a steal phenomenon [3, 4]. Because of the rare condition of all-portal system occlusion with inferior mesenteric AVM, severe portal hypertension may have led to refractory esophageal varices in this case.

The treatment options for this condition reported in the literature are surgical shunt ligation, surgical shunt resection, TAE, transvenous embolization (TVE), intestinal resection for ischemic bowel disease, and combined procedures [1, 3-5, 8]. In most cases of ischemic colitis, intestinal resection is initial or secondary, mostly after the failure of TAE [1, 3-5]. Only one report of ischemic bowel disease involved successful TAE of an identified AVM using an Onyx liquid embolic agent without intestinal resection [6]. TAE is considered ineffective or avoided if it could achieve complete occlusion of shunt points in cases of multiple fine inflow arteries. Moreover, TAE entails a risk of causing ischemic bowel disease by embolizing feeders to the intestines or when embolic material passes into the portal circulation [1, 5]. In particular, migration of embolic material can occur when the diameter of the shunt vessel is greater than 8 mm and has a high flow rate [3]. In the current case, we selected TAE because the shunts were less than 8 mm in diameter. For embolic materials, liquid embolization material, for example, n-butyl-2-cyanoacrylate with flow control, was the best to achieve total nidus vascular bed occlusion. However, using liquid material has the risk of causing liver dysfunction because inflow to the liver is maintained by very fine collateral vessels from the partially patent SMV. Therefore, we chose to use metallic coils because inflow to the liver is maintained by very fine collateral vessels from the partially patent SMV and the migrated liquid embolic material through the nidus might cause the flow impairment of such fine collaterals. In addition, proximal embolization and the development of new collateral nidus-like vessels were permitted. In the past, two patients presented with esophageal varices: one was surgically treated after failure of endoscopic treatment [5], and the other was treated by surgical splenorenal shunt plasty, followed by TVE [8]. The advantage of TVE over TAE is the achievement of complete embolization of the outflow vein without causing ischemic bowel disease. However, TVE requires percutaneous portal system puncture, followed by minor laparotomy and direct exposure of the targeted vessels, which is a risk factor for peritoneal bleeding. In the present case, TVE was not considered an indication because the portal vein was occluded.

Even if direct puncture of the dilated marginal vein was achieved, embolization of such wide and high-flow vessels would have been technically difficult.

The most notable feature of the current case was the total occlusion of all portal systems. To date, there have been no reports of inferior mesenteric AVFs/AVMs with total occlusion of all portal systems. This finding may be associated with idiopathic myointimal hyperplasia of the IMV (IMHMV). Shah et al. reported a case of inferior mesenteric AVFs treated with TAE, followed by colonic resection. Pathological analysis revealed prominent concentric intimal smooth muscle hyperplasia and colonic perforations. They concluded that IMHMV was caused by inferior mesenteric AVFs and that previous IMHMV reports may have been associated with potential inferior mesenteric AVFs [7]. Guadagno et al. and Hendy et al. [6, 9] reported similar results. On the basis of these reports, we hypothesized that continuous arterial pressure to the portal system since birth caused the myointimal hyperplasia of the IMV, splenic vein, SMV, and portal vein in the current case. Because of the progression of myointimal hyperplasia over time, thrombotic occlusion of all portal systems occurred.

The origin of AVM in this case is controversial. Because this patient had no history of abdominal surgery or trauma, this case was initially thought to be congenital. However, not only the genetic factor but also the environmental factors such as hypoxic insult, radiation exposure, epilepsy, and inflammatory process play an important role in forming AVM according to the recent expert's consensus [10]. Thus, it was difficult to determine whether this case was congenital.

In conclusion, we presented a rare case of refractory esophageal varices caused by an inferior mesenteric AVM with all-portal system occlusion that was successfully treated with TAE.

Conflict of Interest: None

Author Contribution: Natsuhiko Saito, Masayoshi Inoue, Hidehiko Taguchi, Junko Takahama: Contributions to the submitted work: the conception or design of the work
Masayoshi Inoue, Kentaro Ishida, Hidehiko Taguchi, Masayo Haga, Emiko Shimoda, Kengo Morimoto, Toshihiro Tanaka, Junko Takahama: Contributions to the submitted work: the advice and review to make manuscript

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