

# Embolization of a duodenal arteriovenous malformation in hereditary hemorrhagic telangiectasia: Case report and review of the literature

Phil Inouye MD MSc<sup>1</sup>, Norman Marcon MD FRCPC<sup>2</sup>, Robin A Pugash MD FRCPC<sup>3</sup>,  
Robert H Hyland MD FRCPC<sup>4</sup>, Marie E Faughnan MD MSc FRCPC<sup>4,5</sup>

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P Inouye, N Marcon, RA Pugash, RH Hyland, ME Faughnan. Embolization of a duodenal arteriovenous malformation in hereditary hemorrhagic telangiectasia: Case report and review of the literature. *Can J Gastroenterol* 2003;17(11):661-665.

A 68-year-old man with hereditary hemorrhagic telangiectasia presented with recurrent, intermittent gastrointestinal hemorrhage. Transfusion of a total of 27 units of red blood cells was required over the three months before admission. Upper and lower endoscopy did not reveal a source of bleeding and a technetium-labelled red blood cell scan was noncontributory. Angiography demonstrated a duodenal arteriovenous malformation originating from a superior mesenteric artery branch. Embolization of the arteriovenous malformation was performed with resolution of gastrointestinal hemorrhage and reduced requirement for blood transfusion. The utility of transcatheter embolization in the management of duodenal arteriovenous malformations in hereditary hemorrhagic telangiectasia is discussed.

**Key Words:** *Arteriovenous malformation; Hereditary hemorrhagic telangiectasia; Gastrointestinal hemorrhage*

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## CASE PRESENTATION

A 68-year-old man with hereditary hemorrhagic telangiectasia (HHT) presented to hospital with recurrent gastrointestinal hemorrhage (GIH), manifesting as intermittent melena, for five years. Previous investigation by esophagogastroduodenoscopy (EGD) revealed four telangiectasias in the stomach that were cauterized. Colonoscopy did not reveal the source of bleeding. The symptoms had progressed, and during the three months before admission to our hospital he had required transfusions of 27 units of packed red blood cells.

Other manifestations of HHT in this patient were moderately severe epistaxis, cutaneous telangiectasias (face, tongue, lips, ear and hands) and bilateral pulmonary arteriovenous malformations (AVMs) for which he had undergone embolization 16 months before the current admission. He also had a history of seizures and transient ischemic attacks; the former were well controlled medically and the latter had resolved after embolization of the pulmonary AVMs. There were no cerebral AVMs. Other past medical history included a left

## L'embolisation de la malformation artérioveineuse duodénale en cas de télangiectasie hémorragique héréditaire : Rapport de cas et analyse bibliographique

Un homme de 68 ans atteint d'une télangiectasie hémorragique héréditaire s'est présenté avec une hémorragie gastro-intestinale récurrente et intermittente. On lui avait transfusé un total de 27 unités de globules rouges au cours des trois mois précédant son admission. Une endoscopie supérieure et inférieure n'a pas permis de révéler la source de l'hémorragie, et une scintigraphie des érythrocytes marqués par technétium n'a rien apporté de plus. L'angiographie a révélé une malformation artérioveineuse du duodénum provenant de l'artère mésentérique supérieure. Une embolisation de la malformation artérioveineuse a été effectuée et a permis de résoudre l'hémorragie gastro-intestinale et de réduire les besoins de transfusion sanguine. L'utilité de l'embolisation transcathétérienne dans la prise en charge des malformations artérioveineuses du duodénum en cas de télangiectasie hémorragique héréditaire est abordée.

renal oncocytoma, hypertension, right cataract and corneal surgery, and tonsillectomy.

The following investigations were completed to identify a source of the bleeding. A technetium-labelled red blood cell (RBC) scan was noncontributory for a site of blood loss. EGD revealed only small telangiectasias in the posterior wall of the stomach. These were not actively bleeding and not thought to be responsible for the clinical presentation of sudden, large volume GIH in this patient. Hemoglobin concentration was 85 g/L on the day of the EGD (normal 130 g/L to 170 g/L). Enteroscopy showed no obvious lesions in the duodenum and colonoscopy to the cecum revealed only small diverticulae but no active bleeding and no clear source of recurrent hemorrhage.

Ultrasound investigation of the abdomen identified a mildly inhomogeneous echo texture of the liver and a solid mass in mid pole of the left kidney consistent with a previously known renal oncocytoma. The gallbladder, common bile duct, pancreas, spleen and great vessels were normal. Mesenteric duplex ultrasound demonstrated markedly elevated flow within the

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<sup>1</sup>Department of Medicine, University of Toronto; <sup>2</sup>Division of Gastroenterology, Department of Medicine, St Michael's Hospital, University of Toronto; <sup>3</sup>Department of Medical Imaging, University of Toronto, Department of Medical Imaging, Sunnybrook and Women's College Health Sciences Centre; <sup>4</sup>Division of Respiratory Medicine, Department of Medicine, St Michael's Hospital, University of Toronto; <sup>5</sup>Financial Support, Squires Club, Nelson Arthur Hyland Foundation, Sonor Foundation, St Michael's Hospital Research Institute, Toronto, Ontario  
Correspondence and reprints: Dr Marie E Faughnan, St Michael's Hospital, 30 Bond Street, Suite 6045, Toronto, Ontario M5B 1W8.  
Telephone 416-864-5516, fax 416-864-5649, e-mail faughnanm@smh.toronto.on.ca  
Received for publication February 18, 2003. Accepted August 20, 2003



**Figure 1)** Selective celiac arteriogram shows dilation of main branches. Patchy ill-defined opacities throughout the liver are typical telangiectasias

celiac axis at slightly less than 4 L/min. Hepatic artery flow was abnormal, but difficult to accurately quantify. There was normal flow in the superior mesenteric artery (SMA) and in the right main and left portal veins.

A mesenteric angiogram was performed from a right retrograde femoral access initially using a flush catheter for an aortogram, and then a 5 French SOS-Omni catheter for selective exams of the celiac, splenic, common hepatic, proper hepatic, gastroduodenal, superior mesenteric and inferior mesenteric arteries. Injection of the SMA showed a small duodenal AVM. Injection of the common hepatic artery showed patchy opacification of all of the hepatic parenchyma, produced by diffuse small telangiectasias with evidence of arteriportal shunting (Figure 1). In addition, there were pancreatic telangiectasias with shunting, and the celiac trunk (Figure 2), common hepatic and splenic arteries were all dilated. No extravasation of contrast, representing active bleeding, was seen by angiography.

The patient underwent superselective arteriography of the SMA to isolate the AVM (Figure 3). Platinum coil embolotherapy was performed, with total occlusion of the vessel feeding the malformation (Figure 4) and resolution of GIH. There were no complications from the procedure. In the following six months there was no history of recurrent GIH; however, the patient required transfusion with six units of RBCs due to ongoing epistaxis.

## DISCUSSION

HHT (also known as Osler-Weber-Rendu disease) is an autosomal dominant disorder characterized by vascular abnormalities, as recently reviewed (1,2). There are two types of vascular lesions in HHT. The first type is the mucocutaneous telangiectasia. Telangiectasias are small focal dilations of capillaries and postcapillary venules within the mucosa resulting in direct connections between arterioles and venules. When these lesions are found within the gastrointestinal tract they may be termed angiodysplasia. The second type of lesion is the visceral AVM. AVMs are direct connections between arteries and veins that lack intervening capillaries and are larger than telangiectasias or angiodysplasia. In patients with HHT, telangiectasias and/or AVMs may be found in the lungs, brain,



**Figure 2)** Abdominal aortogram shows dilation of the aorta to the level of the celiac trunk (secondary to high flow through celiac territory). The celiac trunk and its branches are all massively dilated

skin, nose, liver and pancreas, as well as throughout the gastrointestinal tract.

Telangiectasias occur in the gastrointestinal tract in about 60% of patients with HHT; AVMs are less common. GIH occurs in 10% to 45% of patients with HHT, commonly originating from telangiectasias and presenting as iron deficiency anemia. Less commonly, HHT patients will have bleeding from AVMs, presenting with sudden, massive hemorrhage (3,4). In a retrospective case series of 28 patients with HHT and GIH, 40% of patients presented with an upper GIH, 10% with a lower GIH and in 50% the bleeding site was indeterminate. EGD was performed on 17 of these patients, and while 70% had typical lesions in the stomach, no duodenal lesions were described (5). Duodenal telangiectasias in HHT have been reported by other authors (6-11). There is only one report (12) describing a duodenal AVM in a patient who may have had HHT, based on a family history of a brother with recurrent GIH attributed to congenital telangiectasias. In non-HHT patients, angiodysplastic lesions are also more commonly reported than AVMs in the duodenum (13-16). In one review of 218 patients with bowel AVMs, only five patients were found to have lesions in the duodenum (17).

Diagnosis of gastrointestinal lesions in HHT can be made by endoscopy, angiography, technetium radionuclide scan or laparotomy. Telangiectasias are usually easy to identify by endoscopy (9) but they may be difficult to see in patients with profound anemia (18). Clinically important submucosal AVMs may not be recognized unless they are actively bleeding at the



**Figure 3)** Selective superior mesenteric arteriogram shows tortuous vessels plus small aneurysms (white arrow) in inferior pancreaticoduodenal territory. The black arrow points to the draining vein of this arteriovenous malformation



**Figure 4)** The superior mesenteric arteriogram performed following coil embolotherapy shows no flow through the arteriovenous malformation

time of endoscopy. Furthermore, the yield may be influenced by the type of endoscopic device used. Traditional end-viewing esophagogastroduodenoscopes (at a length of 110 cm) allow visualization to the distal duodenum, to see beyond that, commercially available enteroscopes (200 cm to 250 cm) may be passed with or without an overtube (19,20). In certain cases a side-viewing duodenoscope would be optimal.

Endoscopy is often the initial procedure for treating bowel lesions in patients with HHT. Telangiectasias can be treated by endoscopic thermocoagulation with either bipolar, laser (potassium titanyl phosphate or neodymium:yttrium-aluminum-garnet) or noncontact with argon plasma coagulation (21,22). In patients with HHT, heater probe, bipolar electrocoagulation and laser have comparable results in controlling acute bleeds from telangiectasias (23).

Medical therapy has a role in the management of GIH in some patients with HHT. Estrogen combined with progesterone has been shown to reduce the transfusion need in patients with bleeding malformations in the gastrointestinal tract, including six patients with HHT (24). However, estrogen can be poorly tolerated in men (6). Case reports of using aminocaproic acid have shown conflicting results in reducing the number of blood transfusions required in patients with HHT (25-27). Surgical management may be required in some patients with HHT to prevent ongoing GIH in refractory cases and in patients with larger AVMs thought to be less amenable to medical therapy or endoscopic management.

Visceral arteriography and embolotherapy have been shown to be effective for the diagnosis and treatment of GIH in non-HHT patients (28,29). The site of hemorrhage can be identified by the location of intraluminal extravasation of contrast material or direct identification of vascular malformations. GIH can then be controlled by the selective infusion of vasoconstrictive drugs such as vasopressin, or by selective embolization with coils, polyvinyl alcohol particles, gelatin sponge or tissue adhesive (30,31).

Potential complications of embolotherapy include those related to angiography (groin hematoma, femoral artery injury or thrombosis, embolism, contrast reactions such as anaphylaxis, and contrast-associated renal failure) or those caused by embolization itself (bowel wall ischemia and infarction). Embolization of nontarget organs is possible but unlikely in the hands of experienced interventional radiologists. Superselective embolization is crucial in minimizing the likelihood of gut ischemia, but may be technically quite challenging. In a series of 40 patients reported by Defreyne et al (29), no bowel complications occurred; however, one partial liver lobe and one partial spleen infarction were noted.

There are no reports of small bowel AVMs treated by embolization in patients with HHT; however, there have been reports in non-HHT patients. In patients undergoing embolization for acute nonvariceal GIH, the bleeding lesion was devascularized by embolization in 39 of 40 patients (29). The most common source of bleeding in these patients were ulcers. In one non-HHT patient, a jejunal AVM was the source



of bleeding and showed intermittent contrast medium extravasation during angiography (28). The jejunal artery was embolized with polyvinyl alcohol and the patient was asymptomatic for four months. However, when melena recurred, angiography was used to demonstrate revascularization of the previously embolized lesion. The microcatheter was then used for methylene blue marking to direct laproscopic resection of the involved jejunum and the patient was free of symptoms at 18 months.

Poon and Poon (16) described a duodenal AVM in a patient who did not have HHT. This lesion was embolized using Gelfoam absorbable gelatin sponge (Pharmacia & Upjohn, USA); however, the lesion continued to bleed and the patient underwent laparotomy and ligation of the gastroduodenal artery. In a review of patients who underwent transcatheter embolic occlusion of the gastroduodenal artery for treatment of duodenal bleeding, Granmayeh et al (32) described one patient who had a vascular abnormality as the cause of GIH. Occlusion with wire coils successfully controlled bleeding in this patient for one year. The patient later re-bleed and was successfully treated with occlusion of collaterals. Furthermore, Palmaz et al (31) described two patients with small bowel AVMs who were successfully treated with selective embolization of jejunal and ileal arteries.

When embolization fails to control GIH, angiography can guide surgical resection of the appropriate segment of bowel (12,14). However, the use of angiography has also been reported to misdirect surgical resection, leading to the removal of nor-

mal bowel (15). Catheter injection of methylene blue can be useful to stain involved regions of bowel for resection. Refractory cases in HHT and non-HHT patients often require eventual surgical management (6-8,12,16,33,34).

In the present patient, endoscopic treatment was not possible because the culprit lesion was not identified from within the lumen of the bowel. This is most likely due to the submucosal location of the lesion, anemia and the absence of active bleeding during investigation. In light of the nature of the lesion identified by angiography, medical therapy would not have been appropriate and did not offer a definitive solution. Furthermore, estrogen-progesterone was considered unacceptable to this patient due to the potential adverse effects. Surgery, although a potentially definitive therapy, is far more invasive than transcatheter embolization.

## CONCLUSIONS

In this patient with a single AVM as the source of recurrent GIH, embolotherapy offered a relatively noninvasive and potentially permanent treatment. Embolization is used for treatment of pulmonary and cerebral AVMs in HHT patients (1); however, to our knowledge, this is the first report of a successful embolization of a duodenal AVM in a patient with HHT. Diagnostic angiography should be considered in HHT patients with sudden, massive or refractory GIH and embolization should be considered in the management of AVMs within the duodenum.

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