Case Report

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Esophageal arteriovenous malformation, a rare cause of significant upper gastrointestinal bleeding: Case report and review of literature

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Summary **Gastrointestinal (GI) arteriovenous malformations (AVMs) are a well-known source of bleeding with colon being the most common site, but they can also occur in rare locations like the esophagus which may present with life threatening bleeding. We report the case of a 51-year-old male with end stage renal disease (ESRD) presenting with hematemesis and acute on chronic anemia. Further investigation showed an esophageal AVM which is an unusual location and it was successfully treated with an endoscopic clip instead of argon plasma coagulation (APC) due to its challenging location and esophageal wall motion from breathing. The patient continued to be asymptomatic without any upper and lower GI bleeding during his 20 months follow up period after the endoscopic management. Review of literature showed only 10 cases of AVMs involving esophagus and the average age of presentation was 52 years with a male predominance. We also provide an overview of those cases in the discussion section below.**

Keywords: Arteriovenous malformation, esophagus, ESRD, upper GI bleeding

1. Introduction

AVMs of the GI tract were first visualized endoscopically in 1939 by Renshaw and in 1945 by Grossman (*1*). The overall prevalence of GI AVMs is unknown. They are more common in patients with certain risk factors like chronic kidney disease (CKD). AVMs are responsible for 2-5% of the cases of upper GI bleeding and 3% of lower GI bleeding (*2,3*). The colon is the most common location (*4*), where they are most often found in the cecum and ascending colon (*5*). The small intestine (jejunum> duodenum> ileum) and stomach are the next most common sites of AVMs in the GI tract (*4*), with esophagus being a rare location.

An analysis of 218 patients with arteriographically documented AVMs by Myers *et al*. reported only one case of esophageal AVM (0.5%) (*4*). Another study evaluating the distribution of GI angioectasias in a western population by Bollinger *et al*. reported that none of the patients in their study had AVMs in the esophagus (*6*). Even though only a handful of cases are available in the literature, it is important to consider esophageal AVMs in the differential diagnosis when a patient presents with an upper GI bleed. We describe a case of significant upper GI bleeding due to an isolated esophageal AVM and its challenges in the endoscopic management.

2. Case Report

A 51-year-old African American man presented to the emergency department (ED) in September of 2016 after having three episodes of gross hematemesis. He described the vomitus as large in quantity with fresh blood and clots. The patient had mild abdominal discomfort before the onset of hematemesis that was

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relieved by vomiting. He denied having melena or hematochezia. He had a history of gastroesophageal reflux disease (GERD), chronic hepatitis B without cirrhosis controlled with lamivudine, hypertension, and ESRD treated with peritoneal dialysis after having a kidney transplant rejection. He denied taking non-steroidal anti-inflammatory drugs (NSAIDs), anticoagulants or antiplatelet drugs. There was no prior history of hematemesis, peptic ulcer disease (PUD), or any known bleeding disorder. An esophagogastroduodenoscopy (EGD), done 7 years before the current admission, was unremarkable, and a colonoscopy, 2 years prior to admission, showed mild colonic diverticulosis.

On admission, the blood pressure was 162/70 mmHg, heart rate was 85 beats/minute, respiration rate was 16 breaths/min and temperature was 36.7°C. The abdominal examination was normal, and the digital rectal exam showed brown stool with a negative occult blood test. Hemoglobin (Hg) 55 g/L with a mean corpuscular volume of 76 FL, white blood cell counts 9.5 × 10^9/L, and platelets 244 × 10^9/L. His baseline Hg prior to this presentation was 9-10 g/dL due to anemia of chronic disease. The last Hg value, two months before the current admission, was 10.3 gm/dL. Prothrombin time and activated partial thromboplastin time were within normal limits. The patient received 1 unit of packed red blood cells (pRBC) transfusion and was started on a continuous infusion of pantoprazole. Hematemesis resolved after the admission, but he required two more units of pRBC to maintain a Hg > 70 g/L. On day two after admission, he had an esophagogastroduodenoscopy (EGD) performed by general surgery for the evaluation of hematemesis which revealed an AVM in the mid-esophagus that was not cauterized due to the location. They recommended the patient be seen by gastroenterology for therapeutic endoscopy. As a complication of the EGD, the patient developed aspiration pneumonia that was evident the day after the procedure, and he was started on antibiotics.

A second EGD procedure was done on day 4 by gastroenterology which revealed a 4 mm mid-esophageal AVM with a small central clean-based ulcer (Figure 1) without active bleeding. Further examination of gastroesophageal junction, gastric and duodenal mucosa was normal. No other source of bleeding was identified. Given the difficult location and esophageal wall motion from breathing, argon plasma coagulation (APC) was not considered to be feasible. A complete obliteration of the AVM was done with a single endoscopic clip. Post application, mucosal anchoring, and obliteration of the lesion was satisfactory (Figure 2). The patient was switched to an oral pantoprazole and was treated for two more days in the hospital for aspiration pneumonia. No further bleeding was reported, Hg remained stable and he was discharged home on histamine H2-receptor antagonists for GERD and oral antibiotics continued for pneumonia. On further follow up at periodic intervals after discharge until May of 2018 the patient reported no further episodes of hematemesis, melena or hematochezia. His Hg returned to his usual of 90-100 g/L.

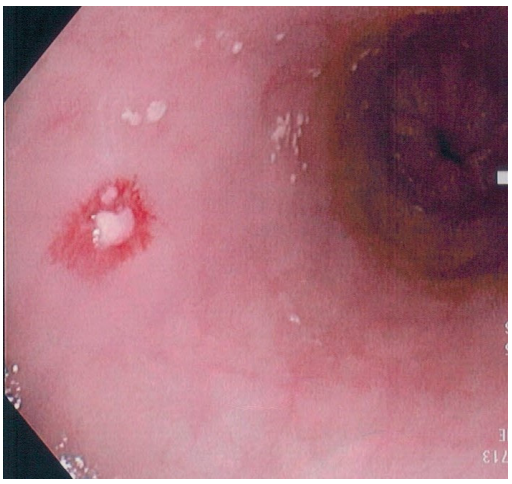

Figure 1. Mid esophageal AVM. A 4 mm mid esophageal arteriovenous malformation with a central clean based ulcer and no stigmata of bleeding at the time of EGD.

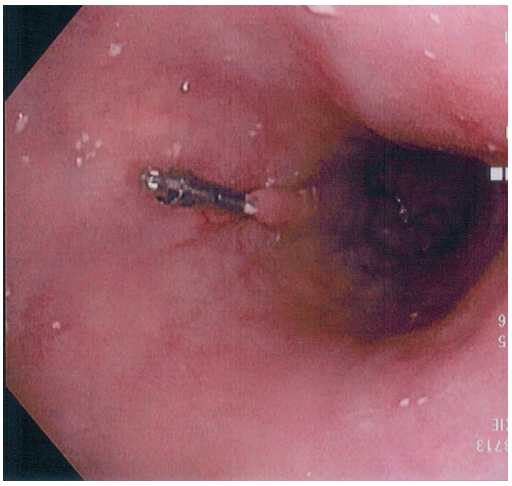

Figure 2. AVM after endoscopic clip placement. A complete obliteration of the mid esophageal arteriovenous malformation after successful application of an endoscopic hemoclip with satisfactory positioning.

3. Discussion

AVMs are associated with various conditions like CKD, aortic stenosis, Von Willebrand's disease, CREST (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) syndrome, and rare conditions like hereditary hemorrhagic telangiectasia (HHT), and Fabry's disease (*3,7*). A study done by Zuckerman *et al*. evaluating the etiology of upper GI bleed (UGIB) showed AVM (53%) as the leading cause in CKD patients and PUD (51%) as the most common cause in patients without renal failure (*8*).

The underlying pathology of AVM formation is

Table 1. Summary of published case reports of patients with AVMs involving the esophagus

Author and year of publication	Patient's age	Sex (M/F)	Anatomical location	Clinical presentation	Treatment modality	Associated conditions	Clinical outcome
Kathi *et al.* (current case)	51	M	Isolated mid esophageal lesion	Hematemesis and anemia	Endoscopic clip	ESRD, GERD, Chronic hepatitis B infection	No recurrence of symptoms after 20 months of follow up
Khanna *et al.* (*14*) (2011)	76	M	Isolated Esophageal lesion	Symptomatic anemia	APC	Gastric Antral Vascular Ectasia (GAVE), Barrett's esophagus, Aortic valve replacement	Not documented
Okano *et al.* (*7*) (2001)	26	M	Esophagus, Angiokeratoma on skin	Hematemesis	Not treated	Fabry's disease	Not treated
Konstantakos *et al.* (*13*) (1995)	69	M	Isolated Esophageal lesion	Dysphagia	Surgery	N/A	No recurrence of symptoms after 6 months of follow up
Kim *et al.* (*16*) (1992)	22	M	Gastroesophageal junction	Hematemesis	Died before treatment and esophageal AVM was found on autopsy	N/A	Death due to hematemesis
Sassaris *et al.* (*15*) (1980)	62	M	Isolated Esophageal lesion	Abdominal pain (antral ulcer noticed on EGD)	N/A	PUD, Chronic pancreatitis	Not documented regarding the treatment for AVM
Sassaris *et al.* (*15*) (1980)	56	F	Esophagus, Stomach	GI bleeding	Endoscopic coagulation attempted but it was unsuccessful	Multiple myeloma, Chronic renal failure	Patient died after massive GI bleed
Weaver *et al.* (*17*) (1979)	71	M	Esophagus, Stomach	Anemia and melena	Endoscopic coagulation	Aortic stenosis, Chronic lung disease, Diverticulosis	Not documented
Schaefer *et al.* (*1*) (1973)	46	M	Esophagus, Stomach	Hematemesis	N/A	HHT	Not documented
Christiansen *et al.* (*18*) (1970)	N/A	N/A	N/A	N/A	N/A	HHT	N/A
Reynolds *et al.* (*19*) (1970)	45	F	Esophagus, Palms, Lips and Tongue	Hematemesis	N/A	CREST syndrome, Primary Biliary Cirrhosis	Not documented

N/A not available.

not well understood. Various hypotheses have been proposed. One suggested possible vascular degeneration promoted by hypo-oxygenation of the mucosa due to atherosclerosis of the vessels (*9*). Other theories suggest increased pressure in the venous system could lead to the formation of the AVMs (*10*).

Clinical presentations of AVM include hematemesis, melena, bleeding per rectum, unexplained iron deficiency and anemia (*4,11*). Diagnosis is usually made by endoscopy, however, in some cases, angiography or surgery may be required to make the diagnosis. Endoscopic therapy with APC is the most successful method of treatment (*12*). Bipolar coagulation can also be used in the treatment of AVM. However, APC is more commonly used due to its ease of use, low cost, and the lower rate of complications. Hemostasis with clips can be used in cases where lesions are localized or APC is difficult to be performed as described in this case.

The PubMed literature search query returned a total of 10 cases of esophageal AVMs reported in combination with AVMs at other locations or as a part of a syndrome (*1,7,13-19*). A brief description of the patients' characteristics of the published case reports is mentioned above in Table 1. Details regarding one of the patients were not available. The most common presentation was hematemesis followed by anemia as it was in the current case. AVMs were seen predominantly in males (8/10) compared to females (2/10) with age ranging from 22-76 years and the average age of presentation was found to be 52. Out of ten patients, only 3 cases of isolated esophageal AVMs were reported (*13-15*), and to the best of our knowledge, the case described here will likely represent the 4th case in that group with its unique challenges in the management and successful hemostasis with an endoscopic clip.

4. Conclusion

Despite its rarity in clinical practice, the case described here provides evidence that esophageal AVMs can cause life threatening upper GI bleeding. They should be considered in the differential in a patient with upper GI bleeding especially in the background of risk factor such as ESRD, and timely intervention could be lifesaving.

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