

CASE REPORT

Calibre persistent submucosal artery of the jejunum: a rare cause of massive gastrointestinal bleeding

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Abstract

Historically a calibre persistent submucosal artery was most often described in the stomach. However in later years it was also discovered in the duodenum and jejunum. It is an uncommon and important cause of massive gastrointestinal bleeding in which failure of detection and early intervention would lead to death. In this paper we report a 27-year-old man with no significant medical history who presented at the emergency unit for recurrent melaena, haematochezia and hypotension. Initial investigations failed to localize the source of bleeding. Emergency exploratory laparotomy revealed a small jejunal mucosal nodule that was actively spurting blood. Histopathological evaluation identified a calibre persistent submucosal artery.

Key words: Persistent calibre submucosal artery, Dieulafoy's lesion, cirroid aneurysm, exulceratio simplex

INTRODUCTION

Persistent calibre submucosal artery or Dieulafoy's lesion is a small, thickened arterial malformation that works its way towards the luminal surface, erodes through the overlying mucosa of the stomach or small intestine, and results in massive arterial haemorrhage.¹ It is characterized by the following: a negative past medical history, independence from peptic ulcer disease, sudden onset, increasing bouts of haematemesis, subcardial location, a tiny mucosal lesion, and open submucosal artery of seemingly large calibre, failure of conservatism in treatment and 60.5 percent overall mortality.² Earlier on, this lesion was only noted in the stomach but in later years it was also found in the duodenum and jejunum.^{3,4,5} There was no histological distinction between gastric and small intestinal cirroid aneurysm.^{4,6,7} This disease poses a diagnostic problem to the surgeon as Dieulafoy's lesion is difficult to diagnose and localize. Error in diagnostic identification is primarily based on the degree of active bleeding at the time of interventional studies and the adequacy of bowel preparation.⁵ In this paper we report a case of Dieulafoy's lesion with Meckel diverticulum that presented with massive lower gastrointestinal bleeding.

CASE REPORT

Mr MRO, a 27-year-old lorry driver who was previously well, presented at the accident and emergency department with complaints of giddiness and fainting episodes. He also had one occasion of passing out melaenic stools mixed with fresh blood just before coming to the hospital. No haematemesis was noted. He had no previous history of similar problems and no history of epigastric pain or discomfort or having haemorrhoids. He was not on any medication such as nonsteroidal anti-inflammatory agents or aspirin products.

Physical examination showed a medium-built gentleman with pallor. Blood pressure was 90/40 mmHg and pulse rate was 100 beats per minute. Per rectal examination showed melaena with fresh blood. Other systems were unremarkable.

During admission, he bled on the bed and the blood pressure progressively dropped. Fluid resuscitation was started and he was given four units of fresh frozen plasma and four units of platelets. Patient was brought to the operation theatre after his vital signs were stabilized. Oesophagogastroduodenoscopy was performed failed to locate the source of bleeding. Emergency exploratory laparotomy was then performed and

a Meckel diverticulum was identified at 20 cm from the ileocaecal junction, however it was not the source of bleeding. Later, a small nodule on the jejunal mucosa located 40 centimeter from duodenojejunal junction was identified through palpation from the serosal aspect. Jejunostomy was performed and revealed an arterial spurting from the nodule. The small and large bowels were filled with fresh and old blood. Jejunal resection with primary anastomosis and Meckel diverticulectomy was performed. Liver, stomach, spleen, large bowel and other parts of the small bowel were unremarkable.

The patient subsequently recovered with no further bleeding during a 2-year follow-up.

Pathology

Macroscopical examination of the resected jejunum showed a cut opened segment of small bowel measuring 5 centimeter in length and 3 centimeter at the largest circumference. On the mucosal surface there was an elevated nodule measuring 0.8 x 0.5 x 0.5 centimeter in diameter. The Meckel diverticulum measured 4 centimeter in length and 2.5 centimeter in the largest diameter. Microscopical examination of the jejunal nodule showed a large submucosal artery with thrombus attached to the wall (Fig.1). On further sectioning, an area of mucosal ulceration communicating with the luminal aspect of the large caliber submucosal artery was noted. No

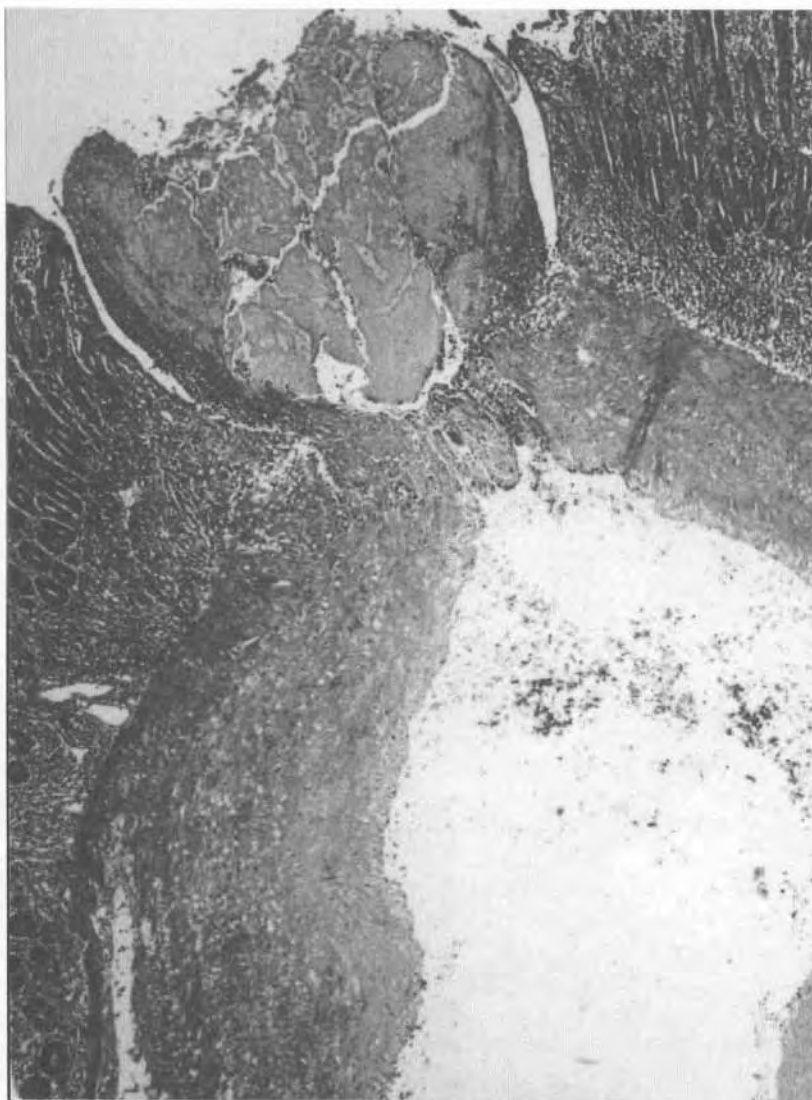


FIG. 1: Large submucosal artery with thrombus attached to the mucosal surface.
H&E X 20.

atherosclerotic changes or vasculitis of the arterial wall was seen. Sections from the Meckel diverticulum showed a lining mucosa of intestinal type but no mucosal ulceration, gastric or pancreatic tissue was seen.

DISCUSSION

Massive upper gastrointestinal bleeding secondary to a dilated submucosal vessel in the stomach was first described by Gallard in 1884.⁸ He gave an account of submucosal miliary aneurysm. After Dieulafoy reported three cases of haemorrhage occurring in young men with this same lesion in 1889,⁹ the disease became known as "exulceratio simplex Dieulafoy" with the belief that this lesion was an early stage of peptic ulceration.^{10,11} There are many other terms used for this lesion including cirroid aneurysm, submucosal arterial malformation,^{12,13} calibre persistent artery of the stomach,^{14,15} gastric arteriosclerosis,¹⁶ and peptic ulcer of peculiar location.¹⁷ The term "cirroid aneurysm" was first coined by Mallory¹⁷ in 1946 to describe the large tortuous vessel contained in the lesion and later were supported by Finkel and Schwartz⁷ as the most appropriate designation. Miko TL in his study excluded any form of aneurysm in this lesion as he proved that the artery was of normal calibre.¹⁴ Recently there has been a consensus that this lesion is a congenital malformation characterized by a single layer, tortuous, and calibre persistent submucosal artery with no evidence of vasculitis, arteriosclerosis, or aneurysm formation.^{10,14} The cause of bleeding is believed to be a mucosal erosion resulting from an artery protruding into the overlying mucosa or from focal gastritis, leading to exposure of the vessel.

Exact data on the incidence of the lesion is rather scarce. Broberg *et al* found three in 210 consecutive cases of massive gastrointestinal bleeding.¹⁸ Strong quoted a prevalence of 0.28 percent of patients presenting with upper gastrointestinal bleeding and it may be as high as 1.3 – 2.3 percent in patients with massive haemorrhage.^{18,19,20,21} A search of the past 30 years of literature yielded four cases of jejunal cirroid aneurysms. The average age at presentation was 44 years.^{4,6,22} There was a male predominance accounting for 86 to 89 percent of the cases in larger series.³

Patients usually had no significant past medical history although mid epigastric complaints were noted for approximately half of the patients.^{4,18,23,24} Its typical clinical

presentation is life threatening massive recurrent gastrointestinal bleeding with anaemia and hypotension. The progression of bleeding cannot be contained by conservative measures, and the only chance for the patient's survival is an operative or some other actively staunching approach.

Diagnosis and localization of the lesion preoperatively is most often unsuccessful. Barium studies have been uniformly not useful in diagnosing this entity.^{4,23,24,25} Endoscopy has been unreliable, as often repeated attempts are required, false negative results are obtained, the lesion is beyond reach, visualization is obscured by blood or ulceration and inflammation may mask the lesion.^{20,25} However endoscopy can at times localize and diagnose the lesion hence some were treated with electrocoagulation.

Results from angiography have been variable in the past. They have been negative in cases where the patient was not actively bleeding.^{4,10,23} Most often the diagnosis was made at exploratory laparotomy. Even with full gastrotomy and duodenotomy may show a normal mucosal surface, and the diagnosis is not made until the mucosa was physically abraded with a dry gauze pad.³

The mainstay of treatment continues to be surgical. Many reports document the success of simple ligation or oversewing of the involved vessel.^{18,19,24,25} Other studies have shown success with local excision or wedge resection without recurrent haemorrhage.^{10,12,21} Consistent with other reported cases of jejunal calibre persistent submucosal artery, our patient had no significant past medical history and bleeding was recurrent and profuse so that the patient developed hypovolaemic shock. Investigation was not helpful until exploratory laparotomy and jejunostomy revealed the lesion. Perhaps, this is the first reported case of Meckel's diverticulum, that can be a source of gastrointestinal haemorrhage associated with jejunal Dieulafoy's lesion. Thorough search for the actual bleeder is mandatory as the presence of Meckel's diverticulum could lead to an erroneous diagnosis and treatment. Jejunal resection was curative as patient had no recurrent bleeding after two years of follow up.

In conclusion, Dieulafoy's lesion of the jejunum is rare and a seldom recognized condition. The presentation could be alarming and the disease should be suspected in a young patient with massive lower gastrointestinal bleeding where endoscopy is negative and angiography suggest a small bowel site.

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