

Case Report Original Research Article

## **Massive obscure GI bleeding from idiopathic jejunal varices identified using single balloon enteroscopy**

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### **Abstract**

**Background:** Obscure gastrointestinal bleeding from idiopathic small bowel varices is both a diagnostic and management challenge for physicians. There are very few cases reported in the literature and there is no consensus on management recommendations.

**Aims:** To present the case of a 34-year-old male patient with bleeding from idiopathic jejunal varices and to review similar cases in the literature.

**Methods:** A case of idiopathic jejunal varices is reported. A literature review was conducted and a total of 25 articles describing idiopathic small bowel varices were identified.

**Results:** Case Report: A 34-year-old gentleman was referred for worsening obscure gastrointestinal bleeding and anemia. Anterograde single balloon enteroscopy revealed several petechial like lesions that were not classic for angiodysplasia. These lesions were initially treated with argon plasma coagulation and clipped, which did not resolve the patient's persistent anemia. No venous abnormalities were identified on computed tomography of the abdomen and pelvis with contrast. The patient underwent an endoscopically assisted exploratory laparoscopy that was converted to a laparotomy upon finding of grossly abnormal distal jejunum. Dilated and tortuous varicosities were identified involving approximately 150 cm of small bowel. It was decided to resect the 40 cm segment of jejunum in which varices were visible endoscopically. There was no evidence of thrombosis in the resected specimen. The patient suffered a pulmonary embolism post-operatively, believed to be provoked by the surgery. The patient has had no re-bleeding 12 months post-resection.

**Literature Review:** Both familial and non-familial accounts of small bowel varices in the absence of a primary cause have been reported in the literature. When supportive therapy is insufficient, the most common treatment modality chosen is surgical resection. Select cases have also demonstrated that sclerotherapy and varix dissection can be used for to treat these lesions.

**Conclusions:** Idiopathic small bowel varices pose both diagnostic and therapeutic challenges for physicians. In the literature, several treatment modalities have been shown to be successful; these include surgical resection, varix dissection and sclerotherapy. There is no consensus on the preferred treatment strategy. This report demonstrates endoscopically assisted surgical resection as a viable management strategy for bleeding of idiopathic small bowel varices, an uncommon cause of occult GI bleeding.

**Keywords:** Single balloon enteroscopy; jejunal varices; case report; GI bleeding

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## Introduction

Bleeding from the small intestine is a relatively uncommon occurrence, accounting for only 5% of all gastrointestinal bleeds.<sup>1</sup> One of the causes of small bowel bleeds are ectopic varices, which are most frequently seen as a result of advanced liver cirrhosis and portal hypertension. Other important risk factors include pancreatitis and thrombosis. Rarely, instances of bleeding from idiopathic small bowel varices have been reported in the absence of any underlying cause. The lack of recognition of these lesions poses a diagnostic dilemma and there are few cases reported in the literature. The rarity of these lesions means that there is little information about their diagnosis and management. The high mortality and morbidity associated with gastrointestinal bleeding underlines the importance of prompt diagnosis and management, even when the cause of bleeding may be obscure. Although mortality rates of upper gastrointestinal bleeds vary between studies, one group demonstrated that the mortality of acute variceal bleeds is as high as 15%.<sup>2</sup> This report examines the diagnosis and treatment of idiopathic small bowel varices in a patient without any significant risk factors.

## Purpose

This article reports the case of a 34-year-old male patient with bleeding from idiopathic jejunal varices. A literature search was conducted examining other instances of idiopathic varices in the small bowel.

## Methods

A literature review was conducted in Pub Med with the following search terms

Varicose vein* OR varix	AND	Small bowel OR Small intestine OR duodenum OR ileum OR jejunum	AND	Gastrointestinal OR GI OR intestinal	AND	Bleed* OR hemorrhage OR melena OR hematemesis OR iron deficiency anemia
Filters : Case report; Meta-analysis; Review; Systematic Reviews				Languages : French; English		

This yielded a total of 355 results. Following initial screening of these results, 97 were marked as potentially relevant. Due to inconsistent search terms, articles included in the references of related articles were also reviewed. This allowed for an increase in the number of related articles

that were identified. A total of 25 articles describing idiopathic varices of the small bowel were found. Inclusion criteria was cases of small bowel varices with no underlying cause. Articles which did not rule out possible underlying causes of small bowel varices were excluded.

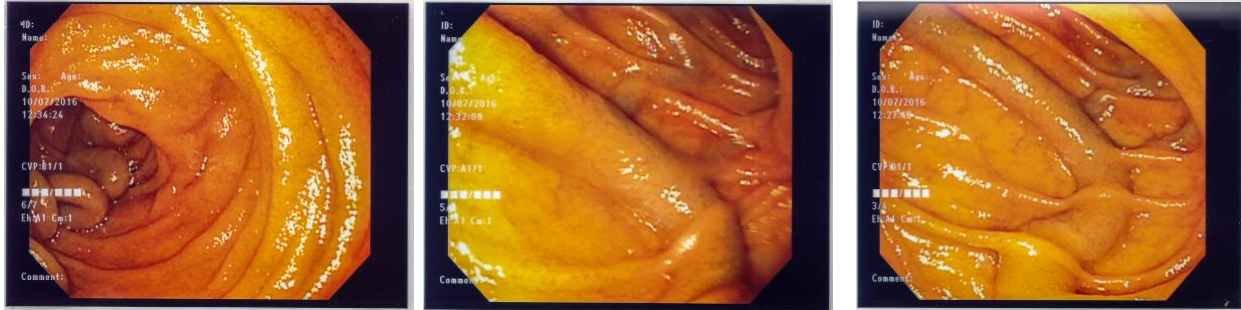
## Case report

A 34-year-old gentleman was referred for worsening obscure gastrointestinal bleeding requiring more than 20 packed red blood cell (pRBC) transfusions over one month. The patient had a history of intermittent melena and anemia over the previous year. He was known for mild asthma and mild developmental delay, but no other significant medical history and specifically no history of liver disease, pancreatitis, or past thrombosis. He presented to emergency with a gastrointestinal bleed, with associated tachycardia and hypotension. Upper and lower endoscopies were non-diagnostic.

On further investigation by capsule endoscopy, he was thought to have potential angiodysplasia of the proximal jejunum. Esophagogastroduodenoscopy (EGD) was unremarkable. Anterograde single balloon enteroscopy revealed several petechial-like lesions that were not classic for angiodysplasia. These lesions were treated with argon plasma coagulation and clipped. However, distal to these lesions in the mid to distal jejunum, large redundant veins were observed although it was not clear over what distance these veins extended. The veins showed no signs of recent bleeding. Small bowel varices were suspected but not treated pending further evaluation. The site was clipped proximal to the veins.

A follow-up computed tomography with contrast of the abdomen and pelvis was performed, and no specific varices or veins were identified. The patient was discharged home with a diagnosis of angiodysplasia bleeding with possible non-bleeding varices.

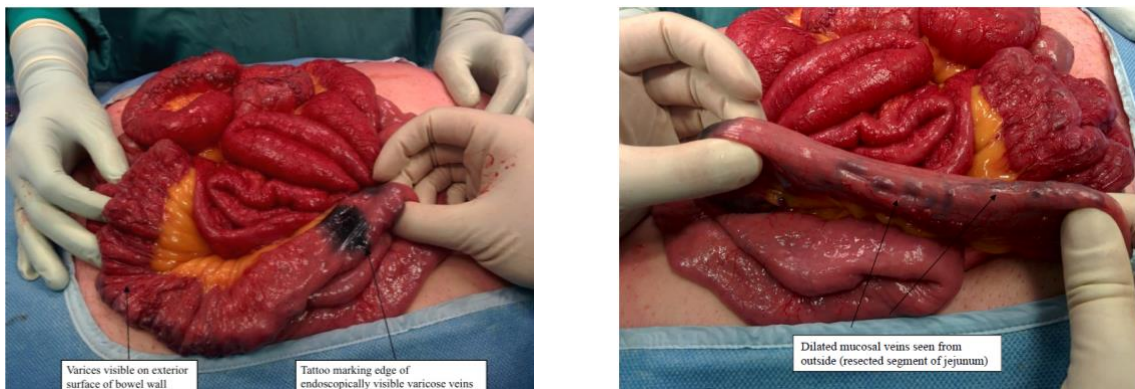
The patient was readmitted following a 3-week follow-up blood test which revealed significant iron deficiency anemia (hemoglobin 75g/L). Two days following the initial follow-up blood test, his anemia at admission had worsened (hemoglobin 69g/L) and he received 2 units of pRBCs. The patient was transferred the following day to a tertiary care center pending intervention and his repeat hemoglobin was 84 g/L. It was assumed that the bleed was likely from the small bowel varices. Patient workup confirmed no evidence of liver disease, portal hypertension, or thrombosis. The single balloon enteroscopy was repeated. There was no evidence of bleeding from the cauterized petechiae or from the visualized varices. (Figure 1)



**Figure 1.** Endoscopy images of the jejunal varices.

Sclerotherapy or histo-acryl glue injection were considered as possible treatment modalities, but it was felt that in the absence of portal hypertension, there was risk of mesenteric and/or portal vein thrombosis. There was concern that there might be further congenital vascular anomalies or hematologic abnormalities that could increase this patient's risk of thrombosis. Furthermore, because the lesion was only visible at the limit of the scope insertion, there was poor control over the needle at the insertion site. As there is no consensus on the preferred treatment modality of jejunal varices in the literature, it was thus decided that the preferred management of the bleeding would be by surgical resection. The site was tattooed.

The patient underwent an endoscopically assisted exploratory laparoscopy that was converted to a laparotomy upon finding of grossly abnormal distal jejunum. The varicose veins extended to just proximal to the ileocecal valve, which was 100 cm proximal to the ligament of Treitz. Interestingly, there were 150 cm of dilated varicosities visible on the exterior of the small bowel, but only 40 cm of the lesion penetrated the submucosa and was visible endoscopically. The colon, mesentery, and mesenteric vasculature appeared normal. The decision was made to resect the 40 cm segment of jejunum for which varices were visible endoscopically. (Figure 2)



**Figure 2.** a) left: Surgical images demonstrating the visibly engorged veins and the endoscopic tattoo b) Right: Dilated veins visible on exterior of bowel wall.

The patient suffered a pulmonary embolism post-operatively 3 days following the resection. The patient presented with hypoxemia and mild chest tightness. Computed tomography pulmonary angiography (CT-PA) confirmed the presence of 2-segmental pulmonary emboli. Following workup and thrombosis consultation, it was determined that this event appeared to be provoked by the surgery. The patient was anticoagulated with Warfarin post-discharge and convalesced without further complications. He was prescribed ferrous fumarate 300mg daily. The patient has had no re-bleeding 8 months post-resection. The patient's hemoglobin increased steadily post-resection and was 107 g/L at his 8-month follow-up appointment. His ferritin remained low at 6ug/L, however, he was no longer experiencing melena stools and had no other evidence of bleeding.

## Discussion

The small bowel is the least common site for gastrointestinal bleeding, however, it is the most common area for bleeds of an obscure nature.<sup>3</sup> The most common site of varices in the small intestine is the duodenum, but lesions may also appear in the jejunum and ileum.<sup>4,5</sup> Since gastrointestinal bleeding is often initially investigated using EGD and colonoscopy, lesions that lie outside the limits of these procedures are missed. In addition, not all small-bowel varices penetrate the submucosa so physicians must be aware that these lesions may not be identified endoscopically.<sup>6</sup> Capsule endoscopy and CT angiography are second line imaging modalities used to identify the source of the bleeding. Specialized endoscopic procedures, such as balloon enteroscopy, can also be used to investigate lesions outside the limits of standard endoscopes. Targeted red blood cell scans have also been used in instances of active bleeding.<sup>7</sup> The success of these new investigative techniques has slowly replaced small bowel radiography.<sup>5</sup>

There is no standardized definition of small bowel varices that occur in the absence of an identifiable underlying cause. The presence of unexplained engorged veins in the small bowel has been described as idiopathic, familial, congenital varices as well as intestinal phlebectasia without an underlying cause. As some authors consider these terms synonymous, we have included all of them in this article. Interventions for ectopic varices can be divided into pharmacological, endoscopic, radiological and surgical options and there is no standard treatment.<sup>8</sup>

A review of the literature identified 25 reports of idiopathic varices in the small intestine. (Table 1) There are reports of both isolated cases and lesions demonstrating evidence of genetic inheritance. (Table 1)

**Table 1.** Reports of Idiopathic Small-bowel Varices.

Case Report	Age at first bleed; Sex	Affected Site	Familial evidence	Treatment	Complications
Atin <i>et al.</i> , 1993 <sup>9</sup>	14 M	Small and large bowel, rectum, oesophagus	Yes	Surgical resection	None

	17 F	Colon, rectosigmoid	Yes	None	N/A
Boland <i>et al.</i> , 2014 <sup>7</sup>	16 M	Large bowel, fundus, proximal stomach	Yes	Supportive, transfusions prn, IV rabeprazole and octreotide	Continued mild bleeding
	13 M	Colon, duodenum, jejunum	Yes	Supportive, transfusions prn, IV rabeprazole and octreotide	None
Dray, X <i>et al.</i> , 2009 <sup>10</sup>	37 F	Colon, ileum	No	Supportive, monthly octreotide acetate LAR injections	None
El-Dosoky <i>et al.</i> , 1994 <sup>11</sup>	10 M	Small and large bowel, rectum, oesophagus	Yes	Past surgical resection	Continued bleeding
Eriguchi <i>et al.</i> , 1998 <sup>12</sup>	49 F	60 cm beyond ligament of Treitz	No	Surgical resection	None
Feldman <i>et al.</i> , 1970 <sup>13</sup>	87 F	Jejunum	Unknown	Surgical resection	None
Friedman <i>et al.</i> , 1990 <sup>14</sup>	32 M	Jejunum	No	Surgical resection	None
Gentili <i>et al.</i> , 2011 <sup>15</sup>	20 F	large bowel, ileum	No	Surgical resection	None
Hanipah <i>et al.</i> , 2016 <sup>8</sup>	61 F	Duodenum	Unknown	Endoscopic sclerotherapy histo-acryl blue	None
Kay & Wylie, 2006 <sup>16</sup>	2 M	Colon, ileum	Suspected	None	N/A
Konishi <i>et al.</i> , 2010 <sup>17</sup>	54 F	Ileum	Unknown	Surgical resection	None
Kumar <i>et al.</i> 1997 <sup>18</sup>	75 F	Jejunum, esophagus	No	None	N/A
Lopes <i>et al.</i> , 2006 <sup>19</sup>	64 M	rectosigmoid, ileum	No	Surgical resection	None
Mejia <i>et al.</i> , 1996 <sup>20</sup>	26 F	Jejunum	No	Surgical resection	None
Morini <i>et al.</i> , 1993 <sup>21</sup>	67 F	Colon, ileum, duodenum	Yes	Unknown	Unknown
	65 F	Colon, suspected small bowel	Yes	Unknown	Unknown
Ohmiya <i>et al.</i> , 2013 <sup>22</sup>	58 M	Jejunum, oral cavity	Unknown	Endoscopic sclerotherapy polidocanol	None, repeat sclerotherapy required
Olano <i>et al.</i> , 2014 <sup>23</sup>	80 F	Jejunum, ileum	Unknown	Conservative therapy, oral iron	None
	66 M	Jejunum. ileum	Unknown	Conservative therapy	Intermittent mild melena
Papnikolau <i>et al.</i> , 2012 <sup>24</sup>	61 M	Ileum	Unknown	Endoscopic sclerotherapy histo-acryl blue	None
Patel <i>et al.</i> , 2008 <sup>25</sup>	14 F	Jejunum	Unknown	Polypectomy of vessel containing polyp by wedge resection laparotomy	Unknown
Peoples <i>et al.</i> , 1981 <sup>26</sup>	60 M	Jejunum, ileum	No	Conservative therapy, iron PO	Unknown
Richardson <i>et al.</i> , 1976 <sup>27</sup>	56 M	Duodenum	Unknown	Varix dissection	None
Rudzki <i>et al.</i> , 2013 <sup>6</sup>	53 F	Duodenum	Unknown	Surgical resection	Pleural effusion secondary to infection
Schilling <i>et al.</i> , 1996 <sup>28</sup>	25 M	Colon, Ileum	No	None	N/A
Tang <i>et al.</i> , 2004 <sup>29</sup>	62 F	Colon, duodenum	No	Conservative therapy, iron PO	N/A
Veillemin <i>et al.</i> , 2004 <sup>30</sup>	34 M	Colon, ileum	No	None	N/A

Surgical resection was the most common treatment modality chosen. Of the surgical cases, 8 had no post-operative complications. However, our patient did suffer a pulmonary embolism post-operatively and there was one reported case of a pleural effusion from a presumed chest infection.<sup>6</sup> It is important to note that there were no reports of massive re-bleeding in any of the patients who underwent surgical resection.

Certain less invasive treatment options have also been explored in the goals of reducing the risk of short bowel syndrome and other complications. Patients have been treated with octreotide acetate long-acting release, which successfully eliminated bleeding of idiopathic varices.<sup>10</sup> Richardson *et al.* reported a case in which variceal bleeding in the second portion of the duodenum was successfully treated by varix dissection from the entrance into the vena cava to its penetration of the duodenal wall.<sup>27</sup> The caval side of the vein was ligated. A full thickness of duodenal wall containing the entire varix was then excised. This patient did well post-operatively and there was no re-bleeding at 8-month follow-up. Endoscopic injection therapy with polidocanol was used to treat varices of the jejunum.<sup>22</sup> Polidocanol is a sclerosing agent that reduces varicosities by causing venous fibrosis. Although the procedure had to be repeated for relapsing phlebectasia, it did ultimately stop the gastrointestinal bleeding. Endoscopic histo-acryl blue injection sclerotherapy has also been used to treat idiopathic varices of the ileum and duodenum.<sup>8,24</sup> This was successful in both reported cases; no complications or occurrences of re-bleeding were reported.<sup>8,24</sup>

Based on the literature, there is no preferred management strategy of idiopathic varices of the small bowel. In patients with high surgical risk but low risk of thrombosis, the less invasive therapies such as sclerotherapy or histo-acryl glue injection may be the preferred management strategies. Furthermore, in patients who have large segments of affected bowel, surgery becomes a less favorable option due to the risk of short-bowel syndrome. However, in cases such as ours where there are suspected underlying venous abnormalities that may increase risk of thrombosis, limitations of endoscopic access to affected area, and relatively short segments of affected bowel, surgical resection becomes advantageous.

## Conclusion

Idiopathic duodenal varices pose both diagnostic and therapeutic challenges for physicians. Presentations of gastrointestinal bleeds caused by these lesions can be acute or chronic. Several treatment modalities have been shown to be successful; these include surgical resection, varix dissection, and sclerotherapy. There is no consensus on the preferred treatment strategy. In the case of our patient, surgical resection of the affected portion of the bowel was effective in achieving hemostasis, however, the patient did suffer a pulmonary embolism post-operatively. This underlines the importance of assessing and managing the risk of thrombosis associated with surgical procedures. It is likely that with the advances of endoscopic techniques which enable



further intubation of the small bowel, such as double balloon endoscopy, these lesions may become easier to investigate.

### Conflict of Interest

The authors have no conflict of interest to disclose.

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