

## **Ectopic Duodenal Varices as a Rare Cause of Bleeding**

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### **ABSTRACT**

**Ectopic varices are dilated veins resulting from portosystemic venous collaterals that appear anywhere in the gastrointestinal mucosa except the gastroesophageal region. Gastrointestinal bleeding caused by this type of varices accounts for 5% of all variceal hemorrhages, making it an unusual cause with a high mortality rate. The optimal treatment for gastrointestinal bleeding due to ectopic duodenal varices has not been well established because of its rarity. Various treatment options have been analyzed for cases with active bleeding, with endoscopic treatment in its various modalities being the preferred choice for achieving initial hemostasis. Additionally, endoscopic treatment can be complemented with therapy guided by interventional radiology, which helps reduce the rate of rebleeding.**

**Keywords:** Duodenal varix, Gastrointestinal bleeding, Variceal bleeding.

### **INTRODUCTION**

Ectopic varices are defined as dilated veins resulting from portosystemic venous collaterals that can appear anywhere in the gastrointestinal mucosa except in the gastroesophageal region. [1,2].

Gastrointestinal bleeding caused by ectopic varices accounts for 5% of all variceal hemorrhages, making it an unusual cause, with a reported mortality rate of 40% during the initial bleeding episode [1,3,4,5].

Up to 40% of patients with intrahepatic portal hypertension develop ectopic duodenal varices, and 10% of them develop anorectal varices [1,6].

Duodenal varices often present with melena or hematochezia and may be associated with hematemesis. In the United States, the most common site for duodenal varices is the duodenal bulb, while in Japan, the second portion of the duodenum is the most common site [7].

In a clinical study conducted in Japan with a total of 173 cases, it was found that the most common afferent vessel for duodenal varices was the inferior pancreaticoduodenal vein in 41%, followed by the superior mesenteric vein. The most frequent efferent vessel was the gonadal vein [8].

The simultaneous presence of esophagogastric varices with ectopic varices is more common in rectal varices than in duodenal varices [8].

The optimal treatment for bleeding duodenal varices has not yet been well established due to their infrequency. However, in cases of acute hemorrhage, urgent endoscopic treatments are preferred [2].

## **MATERIALS AND METHODS**

This study was a multicenter retrospective analysis of individual clinical cases presenting a subset of patients with bleeding ectopic duodenal varices, which were managed with endoscopic treatment. Articles published in indexed international journals from January 2012 to March 2024 were included. Additionally, a case presentation resolved in our medical unit is included.

## **STATISTICAL ANALYSIS**

The cases were evaluated using quantitative variables, expressed as means, and qualitative data, expressed as percentages. The statistical method used for qualitative variables involved 2x2 contingency tables with a cohort design, analyzing differences using Fisher's exact probability test and Chi-squared ( $X^2$ ) analysis.

## **Inclusion and Exclusion Criteria**

Articles with studies and case reports related to ectopic duodenal varices, clinical presentation, treatment, rebleeding rate, and mortality were included. Exclusion criteria consisted of clinical cases presenting anal ectopic varices and those lacking information specified in the inclusion criteria.

## **Results**

A total of 26 cases of duodenal varices were evaluated, of which 15 (58%) were men and 11 (42%) were women. The mean age at presentation was 57.5 years for men and 47.5 years for women.

The most common location of duodenal varices was in the second portion of the duodenum, observed in 11 patients (42.3%), followed by the bulb in 5 patients (19.2%), the third portion in 3 patients (11.5%), the ileum in 1 patient (3.8%), and unspecified portions of the duodenum in 6 patients (23%). (Table 1)

**Table 1: Location of Ectopic Duodenal Varices**

Location	Cases (n)	(%)
<b>Total</b>	26	
<b>Duodenal bulb</b>	5	19.23
<b>Second portion of duodenum</b>	11	42.31
<b>Third portion of duodenum</b>	3	11.54
<b>Ileum</b>	1	3.85
<b>Unspecified</b>	6	23.08

The most common associated comorbidity was alcoholic liver cirrhosis, reported in 9 cases (35%), followed by neoplasms (15%), viral cirrhosis (12%), thrombotic events (12%), and portal hypertension (8%). (Table 2)

**Table 2: Associated Comorbidities in Patients with Ectopic Varices**

Comorbidity	Cases (n)	(%)
<b>Alcoholic liver cirrhosis</b>	9	35
<b>Viral cirrhosis</b>	3	12
<b>Thrombotic events</b>	3	12
<b>Portal hypertension</b>	2	8
<b>Diabetes Mellitus</b>	1	4
<b>Systemic Arterial Hypertension</b>	1	4
<b>Neoplasms</b>	4	15
<b>Unspecified</b>	4	15

The most frequently used endoscopic treatment was the injection of N-butyl-2-cyanoacrylate in combination with another endoscopic therapy, such as band ligation or even with interventional-guided therapies, applied in 45.8% of cases. This was followed by the injection of N-butyl-2-cyanoacrylate as a standalone treatment in 25% of cases. Thirdly, band ligation was used in 12.5% of cases.

Among the 26 isolated cases reported, adequate initial hemostasis was achieved in 24 of them, while 2 patients died due to hypovolemic shock. The overall mortality rate was 19.2%, occurring within the first hours or days following treatment with duodenectomy, coil embolization, N-butyl-2-cyanoacrylate, band ligation, transjugular intrahepatic portosystemic shunt (TIPS), or a combination of these. Adverse effects were observed in 5 patients (15.3%), including bacteremia secondary to the injection of N-butyl-2-cyanoacrylate and a duodenal perforation reported during combined treatment with band ligation, embolization, and sclerotherapy. The main cause of variceal bleeding is liver cirrhosis, which is why patients with duodenal varices may also present with esophagogastric varices. Eleven (42.3%) patients had both esophageal and duodenal varices concurrently. (Table 3)

**Table 3: Treatments Used for Bleeding Ectopic Duodenal Varices and Outcomes**

	Total N= 26 (%)	EBL n= 3	CA n=6	E.Coil n= 2	TIPS n= 1	HC n=1	Combined n= 13	p
<b>Initial Hemostasis</b>	24 (92.3)	3	6	2	1	1	11	NS
<b>Treatment Success</b>	21 (80.7)	3	5	2	1	1	9	NS
<b>Rebleeding</b>	5 (19.2)	0	1	0	0	0	4	NS

<b>Adverse Events</b>	4 (15.3)	0	2	0	0	0	2	NS
<b>Mortality</b>	5 (19.2)	0	1	0	0	0	4	NS
<b>History of Esophagogastric Varices</b>	11 (42.3)	0	4	0	1	1	5	0.04
EBL: Endoscopic band ligation; CA: N-butyl-2-cyanoacrylate; E. Coil: Coil Embolization; TIPS: Transjugular Intrahepatic Portosystemic Shunt; HC: Hemoclip; NS: Not Significant.								

Rebleeding occurred in 5 patients (19.2%), with symptoms such as hematemesis, hematochezia, or melena reported in the first few days following the initial treatment. Among these cases, one patient received treatment with N-butyl-2-cyanoacrylate, achieving adequate hemostasis, while the others required combined treatments, both endoscopic and interventional. Ordinal groups were compared in a multivariate analysis, where rebleeding was associated with the severity of the underlying disease and the presence of esophagogastric varices, using Fisher's exact probability test and Chi-square ( $X^2$ ). No statistical difference was found between those with esophagogastric varices and those without, with a p-value of 0.0434.

## Discussion

Duodenal varices are rare, and when they present with active bleeding, endoscopic treatment is preferred to achieve initial hemostasis. There is a wide range of options, among which the most commonly used are N-butyl-2-cyanoacrylate injection, band ligation, and the placement of hemostatic clips. Different adverse effects have been associated with each of these treatments, such as bacteremia, thrombosis, and migration for N-butyl-2-cyanoacrylate, and the risk of submucosal necrosis for band ligation due to the thinner wall of the duodenum compared to the esophagus and stomach. The endoscopic approach can be particularly challenging due to the anatomy of the duodenum, especially when the variceal path is in close proximity to the ampulla [8].

Other options that have shown good results in achieving hemostasis in duodenal varices include coil embolization, transjugular intrahepatic portosystemic shunt (TIPS), and balloon-assisted transvenous obliteration (B-RTO). No treatment has proven to be ideal for this condition; however, endoscopic treatments are the most commonly used.

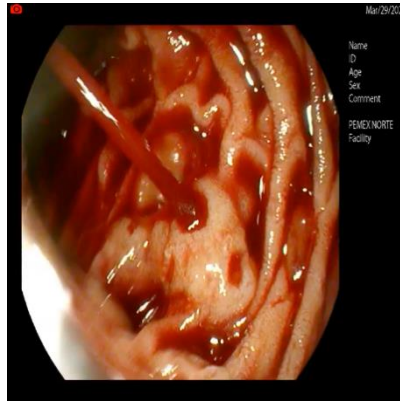
## CASE PRESENTATION

A 73-year-old male was admitted to the emergency department with hypovolemic shock secondary to upper gastrointestinal bleeding, characterized by melena. The patient had a history of alcoholic liver cirrhosis classified as Child-Pugh class A, in addition to hyperuricemia.

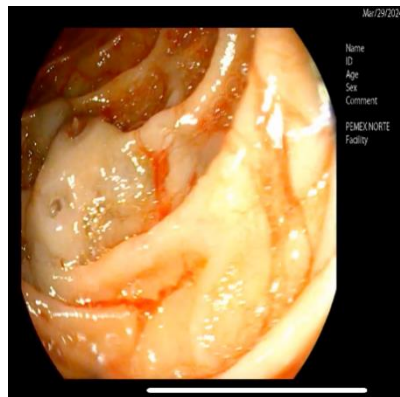
Upon arrival, he presented with hypotension (90/60 mmHg) and tachycardia (110 beats per minute). On physical examination, pallor of the skin was observed, but there was no evidence of ascites. Laboratory results showed a hemoglobin level of 6.7 g/dL, total bilirubin of 0.6 mg/dL, albumin of 3.5 g/dL, and an INR of 1.3, indicating the need for erythrocyte concentrate transfusion.

An urgent esophagogastroduodenoscopy (EGD) was performed, which revealed the presence of an ectopic variceal path in the second portion of the duodenum with active jet bleeding

(Figure 1), which subsided. Two white nipple signs were then observed (Figure 2). Band ligation was performed at both sites, successfully stopping the bleeding (Figure 3). No evidence of esophagogastric varices was found.



**Figure 1: Bleeding duodenal varix**

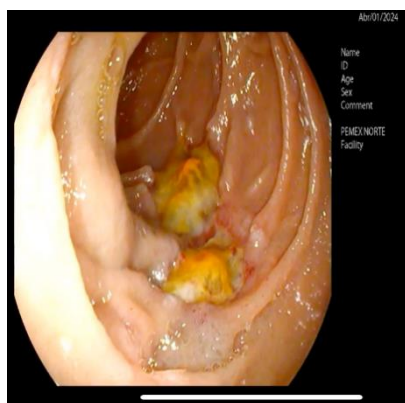


**Figure 2: White nipple sign**

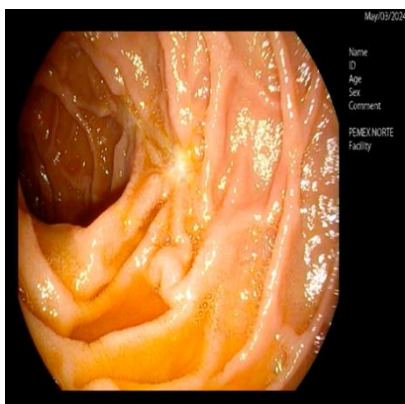


**Figure 3: Band ligation of duodenal variceal tract**

Three days later, a follow-up EGD was performed, which showed superficial ulcers at the band ligation sites, with no signs of bleeding (Figure 4). On follow-up at one month, the patient had no episodes of rebleeding, and a stellate scar was observed at each band ligation site on the follow-up EGD (Figure 5).



**Figure 4: Superficial ulcers covered with fibrin at the band ligation sites**



**Figure 5: Stellate scar in the second portion of the duodenum**

## CONCLUSION

Endoscopic treatment for bleeding duodenal varices has been successful in most cases. Although N-butyl-2-cyanoacrylate has been used more frequently in the cases reported in this study, band ligation has shown good results in the short and medium term. Rebleeding management can be approached with a new EGD or interventional radiology-guided treatment. However, the limited number of cases studied represents a limitation of the study. It is emphasized that the ideal management is achieved through a multidisciplinary approach.

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