

## CASE REPORT

## MODERN MANAGEMENT OF DUODENAL VARICEAL BLEEDING

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Duodenal variceal bleeding is an uncommon cause of gastrointestinal bleeding. Treatment strategies are reliant on case reports and case series with new developments in interventional treatment modalities including endoscopic therapy, radiological intervention, and surgery. Endoscopic treatment includes injection sclerotherapy using various agents, banding of varices and clipping of varices. Interventional radiological procedures include Transjugular Intrahepatic porto-systemic shunt (TIPSS), and Balloon-Occluded Retrograde Transvenous Obliteration (BROTO). Surgical treatment could be suture ligation of varices or gastro-duodenectomy. In this article, a case report of upper gastrointestinal bleed, and management of duodenal varices is described, with review of the literature to offer optimum modern era treatment to these high risk gastrointestinal bleeds.

**Keywords:** Variceal Clips; TIPSS; Ectopic varices; Portal Hypertension; Duodenectomy

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## INTRODUCTION

Duodenal varices are ectopic varices which are described as large portal-systemic venous collaterals occurring anywhere in the abdomen except in the cardio-oesophageal region.<sup>1</sup> Alberti reported the first case of duodenal varices in 1931.<sup>2</sup> The prevalence of ectopic variceal bleeding is 2–5% in patients with liver cirrhosis and between 20–30% of cases with extrahepatic portal hypertension.<sup>3,4</sup> Although, duodenal varices rarely bleed, the mortality is as high as 35–40%.<sup>5,6</sup> Following is a case of duodenal variceal bleeding treated successfully with Transjugular intrahepatic porto-systemic shunt.

## CASE REPORT

A 73-year-old female with background of hypothyroidism, diet controlled type 2 diabetes mellitus and hypertension presented with melaena. She was haemodynamically stable with blood pressure of 110/60 mmHg and heart rate of 90 beats per minute. Her liver function tests including albumin and clotting were normal. An urgent upper gastrointestinal endoscopy showed gastric and duodenal varices figure-1



Figure-1: Duodenal varix seen at OGD

Duodenal varix was the source of bleeding. She was treated with terlipressin, antibiotics, propranolol and blood transfusion in usual manner. A subsequent computed tomography scan showed small, irregular liver with gastric, duodenal, small bowel and sigmoid varices. Spleen was of normal size. A full colonoscopy was performed and ruled out other causes of bleeding. Further evaluation confirmed high alcohol intake as the cause of liver cirrhosis. The liver screen including hepatitis B and C serology, immunoglobulins, liver autoantibody screen and haemochromatosis screen were negative. She was initially transfusion dependent without any overt bleeding but was later discharged home. She was re-admitted the following month with large melaena and was in hypovolaemic shock. She was resuscitated, and then underwent an urgent transjugular intrahepatic porto-systemic shunt (TIPSS) with good result. She had no further bleeding and there were no significant complications including encephalopathy. She has been followed up two years since her TIPSS and has remained well.

## DISCUSSION

Ectopic varices are natural portosystemic shunts due to portal hypertension. Portal venous system is a low resistance system whereas the porto-systemic collaterals have relatively higher resistance and blood favourably flows via the portal system. Normal portal pressure is considered to be 5–10 mmHg. Once the pressure is 12 mmHg or more, complications can arise including ectopic varices. Surgical literature and cadaveric studies have shown following portosystemic communications (a) gastroesophageal plexus to azygous-coronary system, (b) haemorrhoidal plexus, (c) recanalized umbilical vein, and (d) pancreatoduodenal venous arcade to inferior vena cava through retroperitoneal veins of Retzius.<sup>7</sup> These collaterals become patent with increase in

portal pressure usually due to liver cirrhosis and increased intrahepatic vascular resistance. However, extrahepatic portal vein obstruction is well recognised cause of ectopic varices. Surgical procedures with apposition of bowel to abdominal wall or other structures drained by the systemic venous circulation can also cause ectopic varices. The best recognized of these ectopic varices are ileostomy site stomal varices in patients with primary sclerosing cholangitis who had colectomy with ileostomy formation for inflammatory bowel disease, typically ulcerative colitis.<sup>1,8</sup> Even rarer causes are congenital anomalies,<sup>9</sup> arteriovenous fistula,<sup>10</sup> and familial colonic varices<sup>11</sup>. Although, duodenal varices rarely cause gastrointestinal bleeding, there reported angiographic prevalence is up to 40 percent.<sup>3,5</sup> The accepted anatomical reason for this discrepancy is that duodenal varices are usually under serosal surface and in the muscular layers.<sup>12</sup> However, their clinical significance is not apparent until the varix expands into the submucosal space where it can produce gastrointestinal haemorrhage.<sup>6</sup>

Development of duodenal varices could be a result of prolonged portal hypertension or a variation of the same process. Several studies have suggested duodenal varices to be more commonly associated with extrahepatic portal hypertension than intrahepatic causes.<sup>3,6,13</sup> Other mechanisms for duodenal varices include hepatic artery-portal vein fistula<sup>10</sup> and after the loss of oesophageal portosystemic shunts by ablative sclerotherapy<sup>14</sup>.

The treatment of duodenal varices is as varied as its aetiology. Furthermore, factors such as severity of cirrhosis and presence of other ectopic varices dictate the best treatment strategy. Initial management involves haemodynamic stabilisation and correction of clotting abnormalities. Although,

there are no clinical trials for the use of terlipressin or octreotide infusion for duodenal varices they are frequently used. As the above-mentioned drugs reduce portal pressure and have proven benefit in portal hypertension related oesophageal variceal bleeding, their use is perhaps appropriate.<sup>1,15</sup>

More definite treatment depends on available resources and local expertise. The first reported treatment was by Wheeler in 1957 in which his patient underwent suture ligation of the duodenal varices, followed by splenectomy with spleno-renal anastomosis and subsequently internal, external, aneurysmorhaphy and wiring.<sup>10</sup> Nowadays, an upper gastrointestinal endoscopy is the first diagnostic test. Duodenal varix should be considered in the appropriate setting as it can easily be confused with bleeding duodenal ulcer. In fact, one review suggested only 44% endoscopic accuracy preoperatively.<sup>5</sup>

Treatment options available for duodenal varices can be divided in to endoscopic, radiological and surgical techniques.

Endoscopy is the first line as with any case of upper gastrointestinal bleeding with diagnostic and therapeutic value. Endoscopic modalities used in case reports include injection sclerotherapy, band ligation and more recently clipping of the duodenal varix.

Injection sclerotherapy has been used since the beginning of 80's for this condition but the type of sclerosant and volume used has been variable. The type of agents used include ethanolamine, polidocanol, dextrose 50% solution with 3% sodium tetradecylsulfate, thrombin and n-butyl-2-cyanoacrylate.<sup>16-18</sup> As shown in table-1 injection sclerotherapy has been successful with low rates of complications.

**Table-1: Different injectable agents used for bleeding duodenal varices with increasing trend towards N-butyl-2-cyanoacrylate**

Successful injection sclerotherapy for bleeding duodenal varix in intrahepatic portal obstruction	Kirkpatrick R <sup>18</sup>	1985	Fibrin	Successful
Successful endoscopic injection sclerotherapy of a bleeding duodenal varix	Barbish AW <sup>19</sup>	1993	50% dextrose and 3% sodium tetradecyl sulfate	Successful
Treatment of a hemorrhagic duodenal varice by endoscopic sclerotherapy	Paupard T <sup>20</sup>	1995	agent	Successful
Bleeding duodenal varices after gastroesophageal varices ligation: a case report.	Wu WC <sup>21</sup>	2002	N-butyl-2-cyanoacrylate	Successful
Ruptured duodenal varices arising from the main portal vein successfully treated with endoscopic injection sclerotherapy: a case report	Kang <sup>22</sup>	2011	N-butyl-2-cyanoacrylate	Successful
Endoscopic ultrasound-guided treatment of bleeding duodenal varix	Rana SS <sup>23</sup>	2011	N-butyl-2-cyanoacrylate	Successful

Clinicians have been rightfully concerned about risk of perforation in the thin walled duodenum but the cases highlighted in table-1 and a review in 1993 by Barbish and further review in 1995 by Paupard did not substantiate those fears.<sup>19,20</sup> There is growing trend towards N-butyl-2-cyanoacrylate in duodenal varices which has good efficacy for gastric varices already.

Complication rates are low and include re-bleeding, pulmonary embolism and splenic infarction.<sup>19,21</sup> Another very promising agent of current era is thrombin. Along with N-butyl-2-cyanoacrylate it has high haemostatic and low re-bleeding rate.<sup>25</sup> Thrombin is useful alternative with easier administration technique and low complication rates.<sup>26</sup>

Band ligation has also been used since 1995 in several case reports and case series.<sup>27</sup> Although, there are again concerns if the varix is not entirely ligated and the subsequent sloughing will leave a large defect.<sup>1</sup> A recent case series and literature review by Gunerson *et al* shows that about 19 cases of endoscopic band ligation have been reported in the literature since 1995.<sup>28</sup> They are generally safe if used alone or in combination with sclerotherapy. The re-bleeding rates are low and repeat procedures are successful. Only 3 cases out of the 19 reported had re-bleeding. Multiple bands have been used with resultant haemostasis although repeat procedure can be difficult due to fibrosis. In one case, that required repeat ligation by Gunnerson, there was a contained duodenal perforation, but no procedure or bleeding related deaths.

More recently, clipping has also been used successfully, in a single case report as method to treat bleeding duodenal varices.<sup>29</sup>

If endoscopic methods fail then where available, interventional radiology is the next best option. The interventional radiological procedures are classified into two groups: one is for portal-systemic shunt occlusion, the other for portal-systemic shunt creation.<sup>30</sup> Both methods have their advantages and limitations. Portal systemic shunt creation such as Transjugular intrahepatic portosystemic shunt (TIPSS), reduces portal pressure which is the primary mechanism of varices. It is easier to perform than surgical shunts. But because it does not target specific vessels the re-bleed rates are high.<sup>31</sup> Furthermore, there are other limitations, such as significant mortality rate, shunt occlusion, and hepatic encephalopathy.<sup>32,33</sup> Kangawa first reported Balloon-Occluded Retrograde Transvenous Obliteration (BRTO) as an interventional radiological method to treat gastric varices in 1996.<sup>34</sup> This technique has been used to treat about 12 cases of duodenal varices with good haemostasis and low re-bleeding rate, probably due to the more targeted treatment.<sup>30</sup> The only reported complication in the short follow up is induction of oesophageal varices.<sup>30</sup>

Surgery was the first method used in 1957 by Wheeler to treat duodenal varices. They suture ligated the varix, followed by splenectomy with spleno-renal anastomosis and subsequently internal, external, aneurysmorrhaphy and wiring.<sup>10</sup> There have been several case reports treating the varices with suture ligation and also with gastro-duodenectomy.<sup>10,35</sup>

## CONCLUSION

Endoscopy should be the first line method for diagnosis and treatment of duodenal varices. Injection sclerotherapy and endoscopic band ligation

are both relatively safe in treating duodenal varices and provide targeted treatment of the culprit vessel. Repeat endoscopy can be used for re-bleeds. After failure of endoscopic treatment radiological intervention is the second line treatment. Surgery should be reserved where radiology is not available or fails.

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