Japanese Journal of Gastroenterology and Hepatology

Case Report

ISSN: 2435-1210 | Volume 8

Rarely Seen Duodenal Varices Merit Vigilant Endoscopy

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Received: 11 Jun 2022
Accepted: 21 Jun 2022
Published: 27 Jun 2022
J Short Name: JJGH

Keywords:

Endoscopy; Chronic Liver Disease; Duodenal varices; Celiac disease; Serum IgATTG antibodies

Citation:

Malhotra P. Rarely Seen Duodenal Varices Merit Vigilant Endoscopy. J Gstro Hepato. V8(21): 1-3

1. Abstract

1.1. Case Report: We present a thirty year old female who was diagnosed recently to be suffering from cryptogenic related compensated chronic liver disease. She had no history of ascites, pedal edema, upper or lower gastrointestinal bleed or Porto systemic encephalopathy. On evaluation her complete haemogram revealed mild anemia and thrombocytopenia, liver function were mildly deranged with slight increase in serum bilirubin, transaminases and mild coagulopathy as evidenced by International Normalized Ratio (INR) level of 1.3. The renal function test, thyroid profile, blood sugar, serum electrolytes, autoimmune and Wilson profile were normal and viral screen was negative. The ultrasonogram abdomen revealed coarse and shrunken liver with altered echotexture and splenomegaly. The serum IgA TTG (tissue tranglutaminase) antibody test was positive (28.5 I.U./ml). The patient was subjected to upper gastrointestinal endoscopy for screening of varices as well as duodenal biopsies for confirming associated celiac disease. The endoscopy revealed low grade esophageal varices but three prominent duodenal varices located in second part. Normally, duodenal biopsies for confirming celiac disease are also taken from second part of duodenum. In this case a great caution was taken while taking biopsy, so as to avoid trauma to duodenal varices. In such cases even biopsies from first part of duodenum can also be taken.

1.2. Conclusion: A beginner can confuse duodenal varix with mucosal folds or sub mucosal lesion and can attempt unwarranted biopsy that can be life threatening and will require urgent banding or glue injection.

2. Introduction

Duodenal varices are a rare consequence of portal hypertension but can be life threatening, if they bleed. The duodenal varices can be attributed to hepatic (e.g. cirrhosis) or extra hepatic (e.g. portal, splenic or superior mesenteric vein thrombosis) reasons. The esophageal varices are primarily treated with endoscopic variceal ligation (EVL) whereas bleeding gastric fundal varices are usually treated with cyanoacrylate injection or shunt procedures but there is no widely accepted treatment modality for duodenal varices. Duodenal varices represent an ectopic portosystemic shunt [1]. Portosystemic communications in splanchnic hypertension occur through the following routes; i) by way of the gastroesophageal plexus to the azygous system, ii) via the hemorrhoid plexus, iii) via a recannalized umbilical vein and iv) via the pancreatoduodenal venous arcade to the retroperitoneal space to communicate with the inferior vena cava utilizing veins of Retzius [2]. Additionally, surgical or inflammatory adhesions of the intestines act as a route of portosystemic shunting [3]. Although duodenal 'varices as a source of gastrointestinal hemorrhage are rare, representing only one third of all ectopic sources of variceal bleeding, their angiographic prevalence is discordantly high. Ectopic varices are natural large port systemic venous collaterals which appear apart from the gastro esophageal region anywhere in the abdomen [4] and are usually seen in portal hypertension but case reports of familial occurrence in the absence of portal hypertension are there in literature [5, 6].

3. Case Report

A thirty year old female who was diagnosed recently to be suffering from cryptogenic related compensated chronic liver disease. She had

no history of ascites, pedal edema, upper or lower gastrointestinal bleed or portosystemic encephalopathy. On evaluation her complete haemogram revealed mild anemia and thrombocytopenia, liver function were mildly deranged with slight increase in serum bilirubin, transaminases and mild coagulopathy as evidenced by International Normalized Ratio (INR) level of 1.3. The renal function test, thyroid profile, blood sugar, serum electrolytes, autoimmune and Wilson profile were normal and viral screen was negative (Hepatitis A,B,C,E). The ultrasonogram abdomen revealed coarse and shrunken liver with altered echotexture and splenomegaly. The serum IgA TTG (tissue tranglutaminase) antibody test was positive (28.5 I.U./ml). The patient was subjected to upper gastrointestinal endoscopy for screening of varices as well as duodenal biopsies for confirming associated celiac disease. The endoscopy revealed low grade esophageal varices but three prominent duodenal varices located in second part. Normally, duodenal biopsies for confirming celiac disease are also taken from second part of duodenum. In this case a great caution was taken while taking biopsy, so as to avoid trauma to duodenal varices. In such cases even biopsies from first part of duodenum can also be taken.

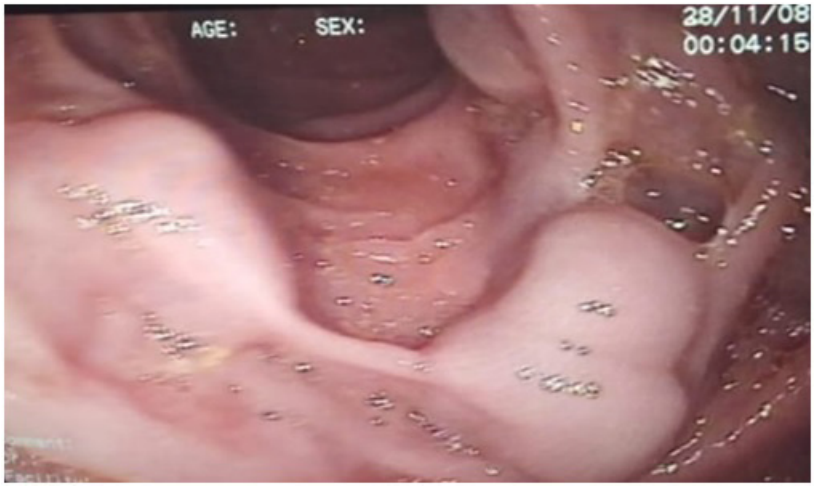

Figure 1: Endoscopy Showing Duodenal Varices Becoming More Prominent in Inhalation.

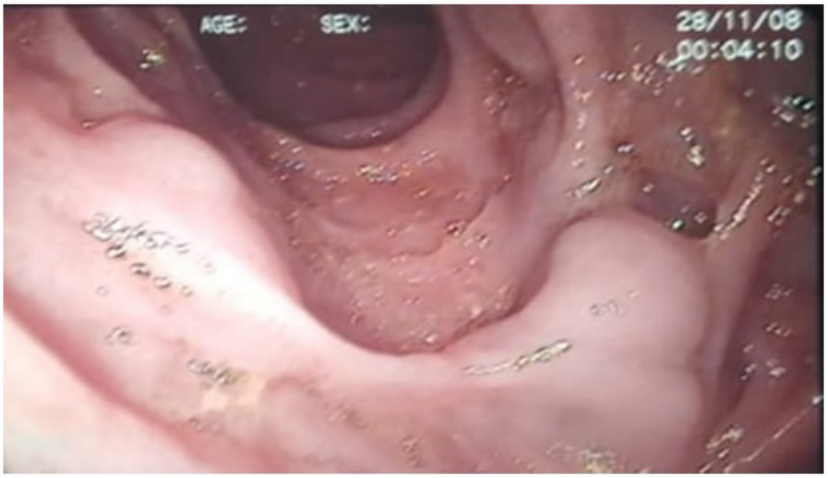

Figure 2: Endoscopy Showing Duodenal Varices Becoming Less Prominent in Exhalation

4. Discussion

The prevalence of ectopic varices varies from 1% to 5% in cirrhotic patients and up to 20% to 30% of patients with extra hepatic portal hypertension (EHPVO). The location of the varices also depends on the cause of portal hypertension [5]. Duodenal varices are found by angiography in more than 40% of patients with EHPVO. Varices in other sites of the small or large intestine are commonly found in patients with cirrhosis, especially in those with history of abdominal surgery, stomas etc. (e.g. patients with primary sclerosing cholangitis who have undergone colectomy and ileostomy for underlying inflammatory bowel disease). The duodenal varices rarely bleed and first report of bleeding from duodenal varices was presented by Alberti et al [7]. The bleeding from duodenal varices can be fatal and mortality rates may reach 35% to 40% [8-10]. The duodenal bulb is the most common location of duodenal varices. Their frequency decreases at the distal duodenum [11]. In contrast to esophageal varices which are sub mucosal, the duodenal varices are usually located in the deeper layers of the duodenal wall. If they are not endoscopically seen, they have no clinical value, since they never bleed. The afferent vessel of the varix is usually the superior or inferior pancreaticoduodenal vein, the superior or inferior mesenteric vein and sometimes the gastroduodenal or pyloric veins [12]. The efferent vein drains into the

inferior vena cava either directly or through the retroperitoneal veins. Duodenal varices seem to have smaller diameter and shorter length than esophageal varices. Wall tension (depending on the vessel size and the portal pressure) seems to be the major determinant of risk of rupture [13]. There have been reports of formation of duodenal varices after injection sclerotherapy or ligation of esophageal or gastric varices [14]. This is probably due to post-treatment alterations in the hemodynamic of portal flow. Management of bleeding duodenal varices is difficult and there are reports of treatment with injection sclerotherapy with different types of sclerosant agents such as ethanolamine, polidocanol, dextrose 50% solution with 3% sodium tetradecylsulfate, polidocanol/thrombin [15, 16]. Emergency sclerotherapy has been shown to be useful as a first-line therapeutic measure in the treatment of bleeding duodenal varices. Endoscopic variceal ligation of ectopic varices has been reported [17], but some authors believe that the banding technique is unsafe for large ectopic varices, since the entire varix cannot be banded and there is also a risk of causing a wide defect in the varix after sloughing off the band [1]. Embolization therapy using radiological techniques is an alternative in the short term management of bleeding ectopic varices and controls bleeding in up to 94% of cases [18, 19]. However rebleeding rates over 1 year are high. In cases of refractory bleed, TIPS or surgical option has to be explored.

5. Conclusion

Endoscopy is an essential test for all chronic liver disease patients but rare presentations like duodenal varix are uncommonly seen. A beginner can confuse duodenal varix with mucosal folds or sub mucosal lesion and can attempt unwarranted biopsy that can be life threatening and will require urgent banding or glue injection. The changes in duodenal varices during different phase of respiration can easily differentiate it with submucosal lesion.

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