

Duodenal polyposis secondary to portal hypertensive duodenopathy

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Abstract

Portal hypertensive duodenopathy (PHD) is a recognized, but uncommon finding of portal hypertension in cirrhotic patients. Lesions associated with PHD include erythema, erosions, ulcers, telangiectasia, exaggerated villous pattern and duodenal varices. However, duodenal polyposis as a manifestation of PHD is rare. We report a case of a 52-year-old man who underwent esophagogastroduodenoscopy and was found with multiple small duodenal polyps ranging in size from 1-8 mm. Biopsy of the representative polyps revealed polypoid fragments of duodenal mucosa with villiform hyperplasia lined by reactive duodenal/gastric foveolar epithelium and underlying lamina propria showed proliferating ectatic and congested capillaries. The features were diagnostic of polyps arising in the setting of PHD.

Key words: Cirrhosis; Portal duodenopathy; Polyposis; Portal hypertension

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Core tip: Duodenal polyposis secondary to portal hypertensive duodenopathy (PHD) is rare. We report a case of PHD presenting as polyposis.

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INTRODUCTION

Portal hypertensive duodenopathy (PHD) is a recognized, but uncommon finding of portal hypertension in cirrhotic patients. While other associations of portal hypertension such as portal hypertensive gastropathy and portal hypertensive colopathy have been described and studied, data concerning duodenal alterations is relatively scarce. The lesions described in PHD include erythema, erosions, ulcers, telangiectasia, exaggerated villous pattern and duodenal varices^[1]. Recently, there have been emerging reports of polyps as a manifestation of PHD^[2-5]. Herein, we report a patient with duodenal polyposis secondary to portal hypertension, review the literature and describe the spectrum of histopathologic changes.

CASE REPORT

A 52-year-old man with compensated alcoholic cirrhosis presented for follow up esophagogastroduodenoscopy. Past medical history includes remote T1N0 colon cancer (status post right hemicolectomy 4 years), low-grade gastrointestinal blood loss, iron deficiency anemia, gastric antral vascular ectasia, portal hypertensive gastropathy and hypertension. He was diagnosed with cirrhosis 13 years ago when he presented with jaundice and ascites and had a recent history of hepatic encephalopathy. Abdominal U/S and magnetic resonance imaging showed a large heterogenous liver, recanalization of the umbilical vein, splenomegaly, splenorenal shunt, additional collateral vessels inferior to the left renal vein and scattered renal cysts. Endoscopy revealed numerous small 1-2 mm polyps extending from the duodenal bulb to the second portion of the duodenum. The three largest polyps included a 6 mm polyp in the mid duodenal bulb (Figures 1A and B), 8 mm polyp distal to this along the anterior wall, and 8 mm polyp in the second part of the duodenum (Figure 1C). The esophagus was normal and no esophageal varices were noted. The stomach showed diffuse "snake skin" appearance, an area of friable mucosa with a polypoid appearance and surface erosions in the antrum and pre-pyloric area with spontaneous oozing of blood. The three duodenal largest polyps were biopsied and histologic examination revealed polypoid fragments of duodenal mucosa with villiform hyperplasia lined by reactive duodenal and gastric foveolar epithelium. The underlying lamina propria showed proliferating ectatic and congested capillaries (Figures 2A and B, D and E). The findings were diagnostic of multiple portal

hypertensive duodenal polyps.

DISCUSSION

Common gastrointestinal tract manifestations of portal hypertension include esophageal/gastric/anorectal varices and gastric antral vascular ectasia. In addition, less common features include portal hypertensive gastropathy^[6-8], congestive jejunosplenic^[9,10], portal colopathy^[11,12] and PHD^[1,13]. PHD is commonly defined as the appearance of patchy or diffuse congestion of the duodenal mucosa associated with friability, erosions or ulcerations^[14,15]. The prevalence of PHD in cirrhotic patients with portal hypertension ranges from 8.4%^[16] to 51.4%^[1]. The lesions described in PHD include erythema, erosions, ulcers, telangiectasia, exaggerated villous pattern and duodenal varices^[1]. Coexistence of severe gastropathy and higher hepatic venous pressure gradients are more frequent in PHD patients and features of PHD have been reported to disappear after liver transplantation^[16].

Duodenal polyps as a manifestation of PHD, an uncommon event, have been reported previously (summarized in Table 1). These include an ulcerated solitary 3 cm polyp in the descending duodenum^[3], multiple sessile polyps in the first portion of the duodenum^[2] and a recent report documenting two to "several" duodenal or jejuno-ileal polypoid lesions ranging in size from < 5 mm to 15 mm in 5 patients^[4]. The spectrum of histopathologic findings in the polyps includes the presence of numerous capillaries with vascular ectasia/congestion/thrombi as well as fibrosis and smooth muscle proliferation. In addition gastric foveolar metaplasia, reactive atypia and ulceration may be seen. Devadason *et al*^[5] reported "duodenal capillary hemangiomas polyps" in 3 pediatric patients (aged 1, 4 and 6 years old). All these 3 patients presented with multiple duodenal polyps in either the 1st or 2nd portion of the duodenum in the setting of extrahepatic portal venous obstruction. Polyps were biopsied in two patients, both of which demonstrated lobular capillary proliferation within the polyps^[5]. Although they favored the term "duodenal capillary hemangiomas polyps", it appears from their description, as well as accompanying image, that the polyps they described share similar morphological features to the polyps in our case and other reported polyps in the setting of PHD.

To date, including our case, there are 11 documented reports of polyps associated with PHD (Table 1). There is no gender predilection (6 male and 5 female), the ages of patients ranges from 1 to 73 years and in the majority of cases (10/11), multiple polyps are seen. The etiology of portal hypertension in adult patients include alcoholic cirrhosis (37.5%, 3/8), hepatitis C cirrhosis (25%, 2/8) and cryptogenic cirrhosis (37.5%, 3/8), while extrahepatic portal venous obstruction accounts for all cases in the pediatric population (100%, 3/3).

Histologically, the PHD associated polyp surface- and crypt-lining epithelium may focally show cells with

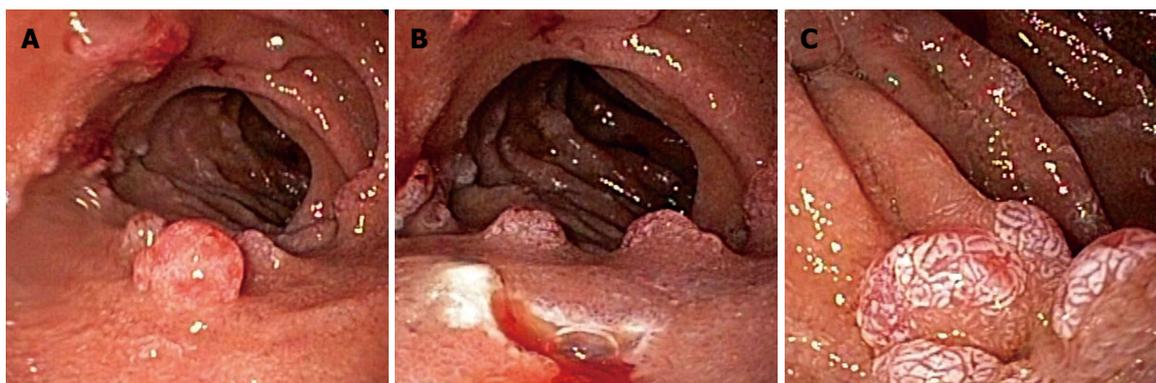


Figure 1 Duodenal polyposis under esophagogastroduodenoscopy. A 6 mm, sessile polyp was seen [prior to removal (A); immediately after removal (B)] in the mid duodenal bulb. A separate 8 mm polyp was seen along the lateral aspect of the second part of the duodenum (C).

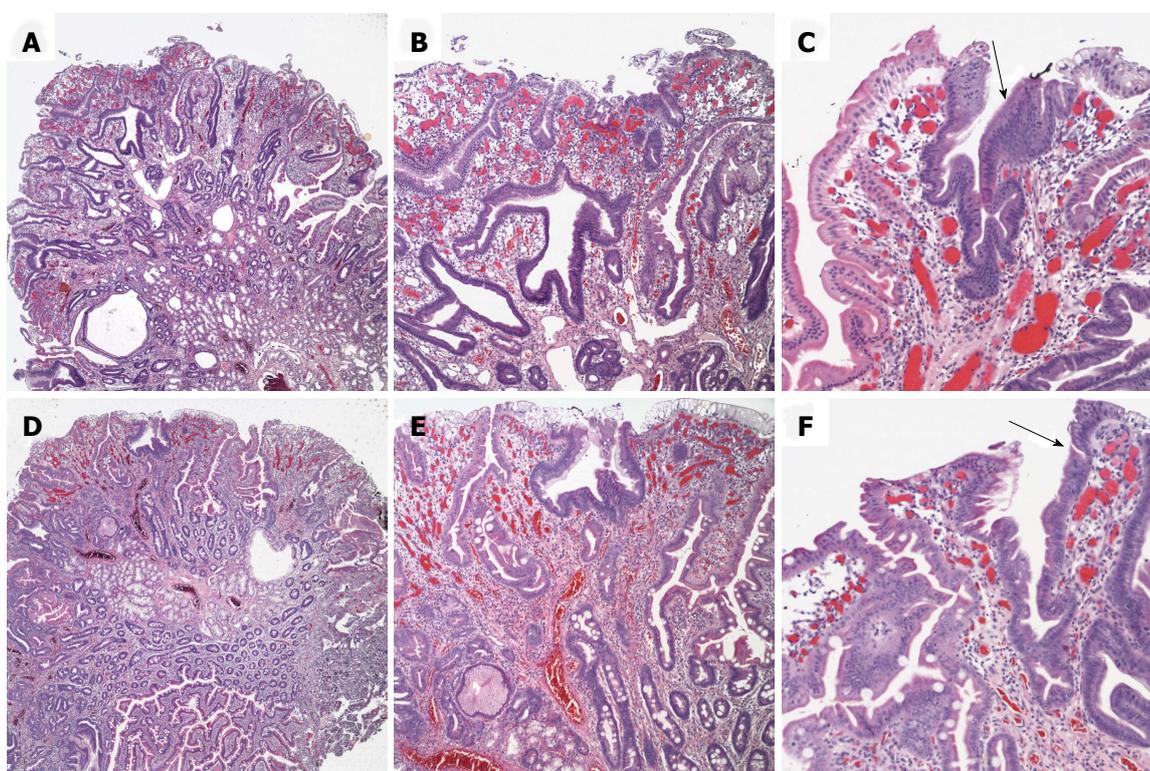


Figure 2 Histopathologic findings. Biopsies from the mid duodenal bulb polyp showed villiform hyperplasia of intestinal and gastric foveolar epithelium with numerous capillaries demonstrating congestion and vascular ectasia (A and B). Similar changes seen in the polyp from the second part of the duodenum (D-E). The epithelium lining the surface and crypts focally (arrows) showing cells with mucin depletion and slightly pencillate nuclei with hyperchromasia (C and F). Representative images of hematoxylin and eosin stained slides taken at 40 × (A, D: 4 × objective and 10 × ocular magnification), 100 × (B, E: 10 × objective and 10 × ocular magnification) and 200 × (C, F: 20 × objective and 10 × ocular magnification).

mucin depletion and contain slightly pencillate nuclei with mild hyperchromasia (Figures 2C and F). These features may mimic duodenal adenomatous polyp, a precancerous lesion in the duodenum. Our current case was previously diagnosed as “duodenal adenomas” at an outside institution. The initial diagnosis of duodenal adenoma in our patient’s prior biopsy highlights the challenges that the reactive atypia may pose during histological evaluation. The differential diagnosis of polypoid lesions in the duodenum is diverse (Table 2) and we limit our discussion to more commonly seen

and lesions with similar histologically features to PHD associated polyps. While duodenal adenomas with low-grade dysplasia (which are histologically similar to those seen in the colon) are typically composed of mucin depleted cells with hyperchromatic pencillate nuclei, compared to the reactive atypia seen in polyps associated with PHD, nuclei show a greater degree of enlargement, hyperchromasia and stratification. PHD polyps differ from duodenal hamartomatous polyps seen in Peutz-Jegher syndrome as polyps in the latter typically show disorganized mucosa with thick arborizing

Table 1 Reported small intestinal polyps secondary to portal hypertension (including current case)

Ref.	Age (yr)/gender	Location(s)	Number/sizes of polyps	Pathologic findings	Etiology of portal hypertension
Current report	52/M	Duodenal bulb to second portion	Greater than 7, majority 1-2 mm, largest 8 mm	Villiform hyperplasia of reactive intestinal and gastric foveolar epithelium, proliferating ectatic and congested lamina propria vessels	Alcoholic cirrhosis
Pillai <i>et al</i> ^[2]	55/M	1 st portion of duodenum	“multiple sessile polyps”, sizes NS	Polypoid muocsa lined by small intestinal and gastric foveolar type epithelium with ectatic capillaries, fibrosis and smooth muscle proliferation of lamina propria	Alcoholic cirrhosis
Zeitoun <i>et al</i> ^[3]	70/M	2 nd portion of duodenum	Single polyp, 3 cm	Numerous thick-walled capillaries with vascular ectasia in lamina propria	Alcoholic cirrhosis
¹ Lemmers <i>et al</i> ^[4]	50/F	Jueuno-ileal	“Several”, > 5 mm	Lamina propria vascular dilation and thrombi without epithelial atypia	Hepatitis C cirrhosis
	73/M	Jejunal	Two “bumps”, < 5 mm	Not biopsied	Cryptogenic cirrhosis
	67/M	Duodenal	“Several”, 5 mm	Lamina propria vascular dilation and inflammation with epithelial atypia and ulceration	Alcoholic cirrhosis
	74/F	Antral/duodenal	“Several”, 15 mm	Lamina propria vascular dilation and epithelium with crenellated glands	Hepatitis C cirrhosis
Devadason <i>et al</i> ^[5]	66/F	Duodenal/jejuno-ileal	“Several”, 5/< 5 mm	Not biopsied	Cryptogenic cirrhosis
	6 yr/M	1 st and 2 nd portion of duodenum	“polyps”, sizes NS	Lobular capillary proliferation in a hemangiomatous pattern in lamina propria	EHPVO
	4 yr/F	2 nd portion of duodenum	“numerous”, sizes NS	Lobular capillary proliferation in a hemangiomatous pattern in lamina propria	EHPVO
	1 yr/F	2 nd portion of duodenum	“polyps”, sizes NS	Polyp not biopsied, mucosa adjacent to polyp with ecatsia and congestion of lamina propria with smooth muscle hypertrophy	EHPVO

¹Data obtained from Table 1 (provided by Dr. Lemmers, personal communication). EHPVO: Extrahepatic portal venous obstruction; NS: Not specified.

Table 2 Histological differential diagnosis of polyps in the duodenum

Primary	Epithelial
	Duodenal adenoma/adenocarcinoma
	Ampullary adenoma/adenocarcinoma
	Hyperplasia, heterotopias, ectopias, inflammatory
	Brunners gland hyperplasia/hamartoma
	Gastric/pancreatic hetertopia/ectopia
	IBD associated inflammatory pseudopolyps
	Inflammatory fibroid polyp
	Peutz Jegher polyps
	Juvenile polyps (JPS or PTEN associated)
	Cronkhite-Canada syndrome polyps
	Neuroendocrine/neural
	Neuroendocrine tumors
	Mixed adenocarcinoma neuroendocrine carcinoma
	Gangliocytic paraglioma
	Neurofibroma
	Ganglioneuroma
	Schwannoma
	Perinerioma
	Mesenchymal
	Gastrointestinal stromal tumor
	Leiomyoma
	Lipoma
	Hemangioma
	Granular cell tumor
	Kaposi sarcoma
	Lymphoid
Lymphoid hyperplasia	
B and T cell lymphomas	
Secondary	Metastases
Miscellaneous	Malakoplakia, mucosal prolapse related, lymphangiectasia, xanthoma

IBD: Inflammatory bowel disease.

smooth muscle fibers of the muscularis mucosa. Although there may be histologic overlap between Juvenile polyps, inflammatory bowel disease (IBD) associated inflammatory polyps and PHD associated polyps, Juvenile polyps are characterized by dilated mucin filled crypts, while IBD associated polyps tend to have prominent glandular architectural distortion in the background of IBD.

In summary, duodenal polyps secondary to PHD is uncommon. With our case, the total number of patients reported in the literature to date is 11. The finding of multiple polyps in a patient with portal hypertension should raise suspicion for this entity and careful histopathologic examination is necessary to render the appropriate diagnosis.

COMMENTS

Case characteristics

A 52-year-old man with compensated alcoholic cirrhosis presented for follow up esophagogastroduodenoscopy and multiple duodenal polyps were found.

Clinical diagnosis

Cirrhosis and duodenal polyps.

Differential diagnosis

Duodenal adenomatous polyp, polyposis syndrome, duodenal pancreatic or gastric ectopia, or other benign neoplasms.

Imaging diagnosis

Endoscopy revealed numerous small 1-2 mm polyps extending from the duodenal bulb to the second portion of the duodenum. The three largest polyps

included a 6 mm polyp in the mid duodenal bulb, 8 mm polyp distal to this along the anterior wall, and 8 mm polyp in the second part of the duodenum.

Pathological diagnosis

Portal hypertensive duodenal polyps.

Related reports

Duodenal polyps as a manifestation of portal hypertensive duodenopathy (PHD), an uncommon event, have been reported previously. The prevalence of PHD in cirrhotic patients with portal hypertension ranges from 8.4% to 51.4%. However, manifestation as multiple duodenal polyps is rare.

Term explanation

Portal hypertensive duodenal polyps are seen in patients with cirrhosis and portal hypertension. The spectrum of histopathologic findings in the polyps includes the presence of numerous capillaries with vascular ectasia/congestion/thrombi as well as fibrosis and smooth muscle proliferation. In addition gastric foveolar metaplasia, reactive atypia and ulceration may be seen.

Experiences and lessons

PHD is a recognized, but uncommon finding of portal hypertension in cirrhotic patients. Multiple duodenal polyps can be an endoscopic finding of PHD.

Peer-review

The authors reported a 52-year-old patient with cirrhosis and portal hypertension who underwent endoscopy and was found with multiple portal hypertensive duodenal polyps. This is an interesting case report and literature review. It is very well written with excellent images. The article highlights the clinical characteristics of PHD and provides information about differential diagnosis of portal hypertensive duodenal polyps.

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