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Portomesenteric venous thrombosis associated with rectal venous malformations☆,☆☆

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Abstract

Purpose: We report thrombosis of portal and mesenteric veins in patients with a pattern of rectal venous malformations (VMs) and ectatic major mesenteric veins.

Methods: Eight patients having rectal VMs with either ectatic mesenteric veins and/or evidence of portomesenteric venous thrombosis (PVT), evaluated from 1995-2009, were reviewed.

Results: Portomesenteric venous thrombosis was evident in 5 patients at presentation. Three had patent ectatic mesenteric veins, 2 with demonstrated reversal of flow, and 2 of whom went on to thrombosis during observation. Six patients developed portal hypertension. Five remain on long-term anticoagulation. After recognizing this pattern, one patient underwent preemptive proximal ligation of the inferior mesenteric vein (IMV) to enhance antegrade portal vein flow and prevent propagation or embolization of venous thrombus from the IMV to the portal vein.

Conclusion: Rectal VMs should be evaluated for associated ectatic mesenteric veins. The ectatic vein siphons flow from the portal vein down to the rectal VM, leading to stagnation of blood in the portal vein and resultant thrombosis. Primary thrombosis in the stagnant rectal VM and/or mesenteric vein can also predispose to embolization up into the portal vein. This pattern of rectal VM and ectatic mesenteric vein should be considered a risk factor for devastating PVT.

Venous malformation (VM), a slow-flow vascular malformation, occurs most commonly in the skin, muscle, and subcutaneous tissues and is therefore usually noted at birth. Occasionally, however, VMs may be encountered in the gastrointestinal (GI) tract. Gastrointestinal VMs may be solitary or multifocal, can involve any or all layers of the bowel wall, and come in a variety of sizes from minute to massive. Although GI VMs may be located anywhere along

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Table 1 Characteristics of patients with rectal VMs

Patient	Sex	Diagnosis VM (age)	Thrombosis (age)	Associated vascular malformation	Ectatic major mesenteric vein or portal trunk
1	F	0	7	KTS	N
2	F	5	7	KTS	N
3	F	0	20	KTS	N
4	M	28	Unknown	KTS	N
5	F	5	–	N	IMV,PV
6	F	12	13	Ileocecal VM	IMV, SMV, PV
7	M	13	13	N	IMV, SMV, PV
8	F	2	18	Sigmoid and hepatic flexure VM	IMV

KTS indicates Klippel-Trenaunay syndrome; Y, yes; N, no; PV, portal vein; SV, splenic vein; SMV, superior mesenteric vein; SRV, superior rectal vein.

the GI tract, most occur as transmural lesions of the left colon and rectum with variable local extension into pelvic structures [1-3]. Although cutaneous VMs are usually reliably diagnosed by history and physical examination, revealing bluish, soft, easily compressible lesions often with palpable phleboliths, rectal VMs may go unnoticed until they manifest with chronic bleeding and anemia [4]. Associated cutaneous vascular anomalies or birthmarks may be present to aid in the initial diagnosis of a rectal VM [4]; however, not all are accompanied by external cues. A thorough history, physical examination, colonoscopy, and appropriate radiologic imaging including contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) to elucidate the location and extent of involvement should be performed when concerned about the presence of a rectal VM. We have recently observed thrombosis of portal and mesenteric veins in patients with a pattern of rectal VM and ectasia of major mesenteric and portal venous trunks. Portomesenteric venous thrombosis (PVT) is potentially lethal and accounts for 5% to 15% of all mesenteric ischemic events in the general population [5]. Patients with PVT will almost always have clinical manifestations of portal hypertension.

1. Materials and methods

Under protocol no. M09-03-0158 issued by the Committee on Clinical Investigation of Children's Hospital Boston (Mass), the Vascular Anomalies Center database was queried for all patients with rectal VMs who presented to our center from 1995 to 2009. Patients were included if they had rectal VM with either ectatic mesenteric veins and/or evidence of PVT. Medical records were reviewed retrospectively for presentation, diagnosis, imaging, treatment modalities, and outcome.

2. Results

A database search identified 26 patients with rectal VMs, 8 of whom were identified as having a rectal VM with either ectatic mesenteric veins and/or evidence of PVT. Among the 8, the median age at time of diagnosis of the rectal VM was 5 years, ranging from 1 week to 28 years (Table 1). Four patients had rectal VMs associated with identifiable ectatic mesenteric or portal venous trunks on imaging. In patients with PVT at presentation, it was not possible to determine whether they had ectatic mesenteric veins previously. Four patients had extensive combined vascular malformations of the Klippel-Trenaunay syndrome. Two patients with rectal VM not associated with Klippel-Trenaunay syndrome had associated colonic and intestinal VMs located in the ileocecal region, sigmoid colon, and hepatic flexure.

Abdominal pain and lower GI bleeding were the 2 most common presenting symptoms and occurred together in 6 patients. The onset of abdominal pain corresponded to the development of PVT in 6 patients. At the time of follow-up, the only patient who has remained free of abdominal pain has not developed PVT. Symptomatic lower GI bleeding was present in all but one patient who had been asymptomatic until presenting with sudden-onset abdominal pain at 13 years of age. Abdominal imaging revealed a rectal VM with ectatic mesenteric veins and portal venous trunk with extensive PVT and hypoperfusion of the right hepatic lobe due thromboembolic occlusion of peripheral branches of the right portal vein.

Portomesenteric venous thrombosis occurred in 7 patients and was present in 5 patients upon presentation. Four patients with Klippel-Trenaunay syndrome and rectal VMs developed PVT. Three of the 4 patients with radiographically documented ectatic mesenteric veins or dilated venous trunks developed PVT. Three patients had patent ectatic mesenteric