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# Imaging Characteristics of Idiopathic Myointimal Hyperplasia of the Mesenteric Veins: Diagnostic Insights

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Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is a rare cause of bowel venous ischemia, which primarily affects the rectosigmoid colon [1,2]. IMHMV presents with a progressive pattern of bowel ischemia characterized by hyperplasia of the intimal smooth muscle of the mesenteric veins, typically involving large-tomedium-sized mesenteric veins. A recent systematic review revealed that 124 cases of IMHMV have been reported in isolated case reports and case series [2] since the initial report in 1991 by Genta et al. [3].

Symptoms of IMHMV include abdominal pain and bloodtinged diarrhea. Endoscopic findings typically include segmental edema, erythema, friability, and multiple ulcers. These clinical and endoscopic findings sometimes overlap with those of inflammatory bowel disease and ischemic colitis. As symptoms often do not resolve easily with medical treatment, patients with IMHMV require repeat colonoscopy and biopsy. Biopsy specimens often show nonspecific signs of ischemic insult, whereas characteristic findings are

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This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (https://creativecommons.org/licenses/by-nc/4.0) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited. typically observed in surgical specimens. Consequently, patients with IMHMV eventually undergo surgery, and definite diagnosis is often delayed.

Considering that the characteristic findings of IMHMV are present in the mesenteric vein rather than in the bowel wall itself, radiological studies could offer valuable insights for early diagnosis. However, due to the rarity of the disease and its similarity to other forms of ischemia, radiologists may not be fully aware of the imaging findings associated with IMHMV. Therefore, it is crucial to be aware of the radiological findings of IMHMV to prevent delays in the diagnosis.

Although clinical and pathological findings have been adequately described in previous studies [2,4], radiological findings have not been well documented in the literature. Therefore, the purpose of this article was to showcase the comprehensive radiologic findings of IMHMV using typical examples and raise awareness of the disease among radiologists.

# **Radiologic Characteristics**

IMHMV is a venous bowel ischemia that progresses at a relatively slow rate. Imaging findings of IMHMV are similar to those observed in other types of venous ischemia. The rectum and sigmoid colon, which are predominantly involved, show marked wall thickening and pericolic fat stranding on computed tomography (CT) and magnetic resonance imaging (MRI). They demonstrate thin highattenuating inner (mucosa) and outer layers (muscularis propria and/or serosa) and thick low-attenuating middle layer (submucosa) on contrast-enhanced CT (Fig. 1). On MRI, pronounced submucosal edema is observed on T2weighted images, whereas diffusion restriction is observed in the mucosal layer and muscularis propria and/or serosa (Fig. 2).

Unlike the mural changes that are similar in other forms of venous bowel ischemia, there are distinct findings in the pericolic vessels that differentiate IMHMV. Notable findings include a slit-like or absent inferior mesenteric vein (IMV), as well as established collateral drainage veins along the proximal colon and mesentery (Fig. 1). These observations may correspond to the pathological characteristics of nonthrombotic and non-inflammatory occlusions of large-to medium-sized mesenteric veins. The presence of collateral draining veins may reflect progressive disease rather than an acute condition. Apparent findings indicating bowel infarction, such as pneumatosis or portomesenteric vein gas, are rare because of the nature of the disease. Additionally, multiple tortuous vessels with attenuation similar to that of the artery and aneurysmal vessels near the affected colon are indicative of IMHMV (Figs. 1, 2). These findings are clearly visible on arterial-dominant

phase images. Therefore, multiphase CT may have additional diagnostic value. Angiography of the mesenteric arteries and subsequent venography are also useful in identifying multiple tortuous artery-like vessels, aneurysmal vessels from the inferior mesenteric artery, dilated collateral veins, and a slit-like or absent IMV (Fig. 1). These findings may be related to the underlying pathogenesis of IMHMV, although this has not been fully elucidated. One plausible theory is that an arteriovenous fistula and persistent venous hypertension result in vascular remodeling, which is hinted at by the pathological similarity between venous changes in IMHMV and those occurring in saphenous vein grafts after coronary artery bypass surgery [5]. An arterialized IMV and its tributaries may become engorged and tortuous, eventually leading to aneurysmal changes.

# **Differential Diagnosis**

IMHMV is often initially misdiagnosed as ulcerative



**Fig. 1.** Imaging findings of a 63-year-old male patient presenting with abdominal pain and blood-tinged diarrhea. **A–C:** CT scans revealing marked thickening of the sigmoid colon (S), exhibiting poor enhancement on the contrast-enhanced images. A slit-like inferior mesenteric vein (IMV) (white arrowheads) appears poorly attenuated. Additionally, tortuous vessels with attenuation similar to that of the artery (white empty arrows) and a small aneurysmal vessel (white empty arrowheads) are visible on CT, CT angiography **(D)**, and inferior mesenteric artery angiography **(E, F). F:** The inferior mesenteric and superior rectal veins are imperceptible, whereas established collateral veins (black arrows) are evident in the delayed angiogram. IMA indicates the origin of the inferior mesenteric artery. CT = computed tomography





**Fig. 2.** Imaging findings of a 79-year-old male patient presenting with abdominal pain and constipation. T2-weighted MR image (**A**, **F**) showing prominent submucosal edema in the sigmoid colon (S) and rectum (R). Diffusion-weighted image (b = 1000) (**B**) and apparent diffusion coefficient map (**C**) demonstrating diffusion restriction in both the mucosa and muscularis propria and/or serosa layers. Thrombosed (white arrowheads) and non-thrombosed (white empty arrowheads) aneurysmal vessels and tortuous artery-like vessels (white empty arrows) are observed near the rectum (**D**-**F**). MR = magnetic resonance

colitis because of the similarities in clinical and endoscopic findings [2]. Ulcerative colitis typically exhibits two age peaks, with the first occurring in the third decade of life and the second between the ages of 40 to 70 years [6,7]. IMHMV has also been reported in healthy middle-aged men [2]. Typical endoscopic findings of ulcerative colitis include edema, erythema, and ulcerations in the left colon and rectum, which are strikingly similar to those observed in IMHMV [4,8]. Severe acute ulcerative colitis shares certain CT and MRI findings with IMHMV, including colon wall thickening with a target sign and prominent vasa recta [9]. However, the presence of multiple tortuous artery-like and aneurysmal vessels around the colon and rectum, which are common findings in IMHMV, are rarely observed in ulcerative colitis. Additionally, the IMV is typically clearly visible in patients with ulcerative colitis, whereas it is absent or severely narrowed in patients with IMHMV.

Ischemic colitis, caused by insufficient arterial supply to the colon rather than venous obstruction, often involves the sigmoid colon and may be confused with IMHMV. The ischemia induces inflammation and subsequent reperfusion, resulting in congestion, edema, and hemorrhage, consequently leading to colon wall thickening, mesenteric infiltration, and ascites [9,10]. Compared with IMHMV, which typically simultaneously affects the sigmoid colon and rectum, rectal involvement is relatively less frequent in ischemic colitis because of the rectum's abundant collateral blood supply [10]. Moreover, a slit-like or occluded IMV is not associated with ischemic colitis.

Acute colon ischemia due to mesenteric venous thrombosis should be distinguished from IMHMV. It presents as luminal filling defects with a thin peripherally enhancing rim corresponding to the venous wall. The tributaries to the occluded vein are occasionally engorged on contrastenhanced CT and MRI [11,12]. By contrast, obliteration of the mesenteric vein without thrombosis is a characteristic feature of IMHMV. Furthermore, mesenteric venous thrombosis does not exhibit tortuous vessels with attenuation, similar to that of the artery and aneurysmal vessels. Another important distinction is that, unlike IMHMV, mesenteric venous



thrombosis is often associated with medical comorbidities that predispose the patient to thrombosis, such as abdominal infection, neoplasm, protein C and/or protein S deficiencies, and oral contraceptive use [13,14].

Severe infectious colitis such as pseudomembranous and cytomegalovirus colitis can produce imaging findings that mimic those of IMHMV. Pseudomembranous colitis involves the colon to varying extents, revealing marked wall thickening and pericolic stranding, and ascites on CT [12,15]. However, the characteristic vascular abnormalities of IMHMV have not been observed in these diseases.

# **CONCLUSION**

IMHMV has distinctive radiological features that allow discrimination from other diseases: 1) marked wall thickening of the rectum and sigmoid colon presenting with reduced enhancement; 2) slit-like or absent IMV with collateral veins along the proximal colon; and 3) multiple pericolic tortuous artery-like vessels with or without aneurysmal vessels. Contrast-enhanced CT scans of the abdomen and pelvis are essential to evaluate mural changes and their extent and the IMV. Multiphase abdominopelvic CT and CT or catheter angiography are also recommended for identifying vascular abnormalities. Therefore, awareness of these findings may facilitate timely diagnosis of IMHMV.

#### **Conflicts of Interest**

The authors have no potential conflicts of interest to disclose.

## **Author Contributions**

Conceptualization: Rohee Park, Jong Keon Jang, Hyun Jin Kim. Formal analysis: Jong Keon Jang. Investigation: Rohee Park, Jong Keon Jang. Methodology: Rohee Park, Jong Keon Jang. Project administration: Jong Keon Jang. Resources: Jihun Kim, Seok-Byung Lim. Supervision: Jong Keon Jang, Hyun Jin Kim, Jihun Kim, Seok-Byung Lim. Visualization: Rohee Park, Jong Keon Jang. Writing—original draft: Rohee Park. Writing—review & editing: Jong Keon Jang, Hyun Jin Kim, Jihun Kim, Seok-Byung Lim.

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