Diffuse cavernous hemangiolymphangioma of the rectosigmoid, diagnosed by contrast-enhanced EUS

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Diffuse cavernous hemangioma (DCH) of the rectosigmoid is a rare congenital venous malformation resulting in hematochezia.[1-5] We report a rare case of DCH associated with lymphangioma, successfully diagnosed by contrast-enhanced EUS.

A 69-year-old woman was admitted for hematochezia and severe anemia. She had a long history of intermittent rectal bleeding since childhood, initially diagnosed as hemorrhoids. These were surgically treated when she was 30 years old. She was then diagnosed with arterial fibrillation at 64 years of age and was administered anticoagulants. Subsequently, her rectal bleeding worsened, requiring blood transfusions. Anticoagulant therapy was stopped after catheter ablation but was resumed upon relapse at 69 years of age. Re-administration of anticoagulants exacerbated her rectal bleeding.

Laboratory findings revealed a hemoglobin level of 6.2 g/dL. Endoscopic examination of the rectosigmoid showed diffuse bluish-purple polyploid lesions and patchy yellowish areas (Fig. 1). Barium contrast study showed narrowing of the distal sigmoid causing luminal filling defects in the rectosigmoid and calcifications in the pelvis (Fig. 2). CT showed diffuse thickening of the rectum and distal sigmoid and multiple pelvic phleboliths within the vascular sinuses in and around the rectum inside and outside the rectal walls (Fig. 3).

EUS revealed a thickened rectosigmoid wall with high echoic content but with obscure layer structures. Small anechoic areas and shadows of calcifications scattered throughout the rectal wall were also observed (Fig. 4A). Doppler imaging did not detect any blood flow.

Contrast-enhanced EUS using perfluorobutane distinctively revealed a minute septum-like structure and slow flow into the small cavities of only the high echoic areas and not of the anechoic areas (Video 1 available online at www.VideoGIE.org, Fig. 4B and C). These parameters are undetectable by conventional EUS and Doppler examinations.

The patient underwent laparoscopic super-low anterior resection. Pathologic examination showed an irregularly dilated vascular overgrowth that filled the submucosal layer and extended into the serosa. The vascular structure consisted of the following 2 components: hemangioma that correlated with the high echoic areas, containing red blood cells lined with CD31-positive cells (Fig. 5A and B), and lymphangioma that correlated with the anechoic areas, containing eosinophilic liquid lined with D2-40–stained endothelial cells (Fig. 5C and D).

The patient was finally diagnosed with diffuse cavernous hemangiolymphangioma of the rectosigmoid. Rectal bleeding stopped and anemia improved after the surgery. This case report thus demonstrated the following 2 important clinical findings: the effectiveness of EUS in diagnosing submucosal lesions, such as DCH, and the usefulness of contrast-enhanced EUS in characterizing minute structures and slow flow hemodynamics of lesions in vivo. Despite its

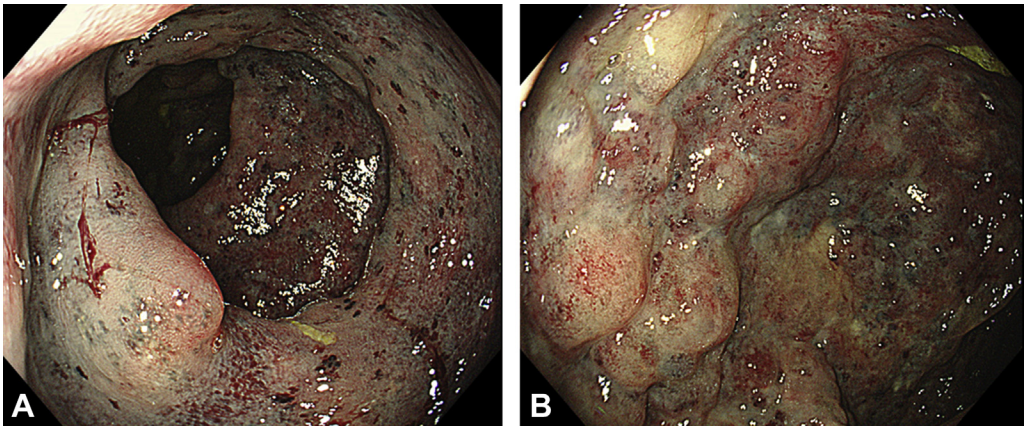

Figure 1. Endoscopic examination showed diffuse bluish-purple polypoid lesions and patchy yellowish areas.

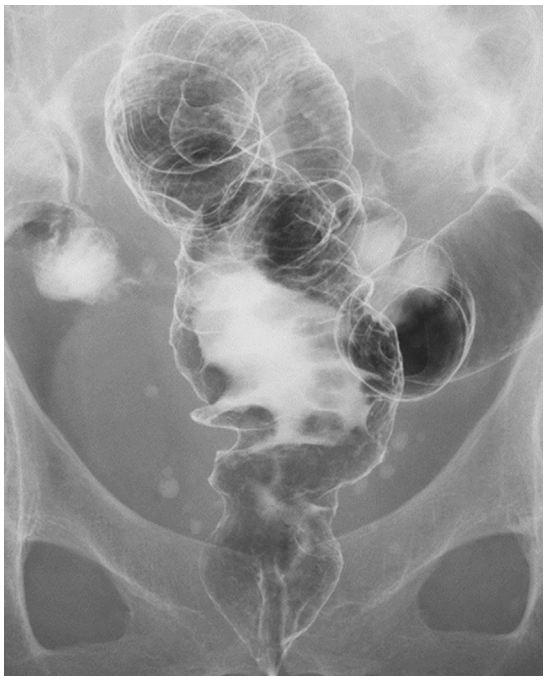

Figure 2. Barium contrast study showed narrowing of the rectosigmoid lumen and luminal filling defects. Calcifications in the pelvis were observed.

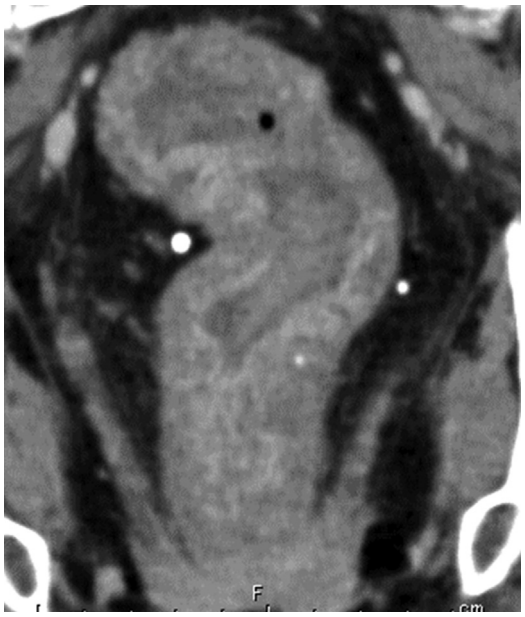

Figure 3. CT showed the thickened rectosigmoid wall to be diffusely enhanced, as well as multiple pelvic phleboliths in and around the rectal wall.

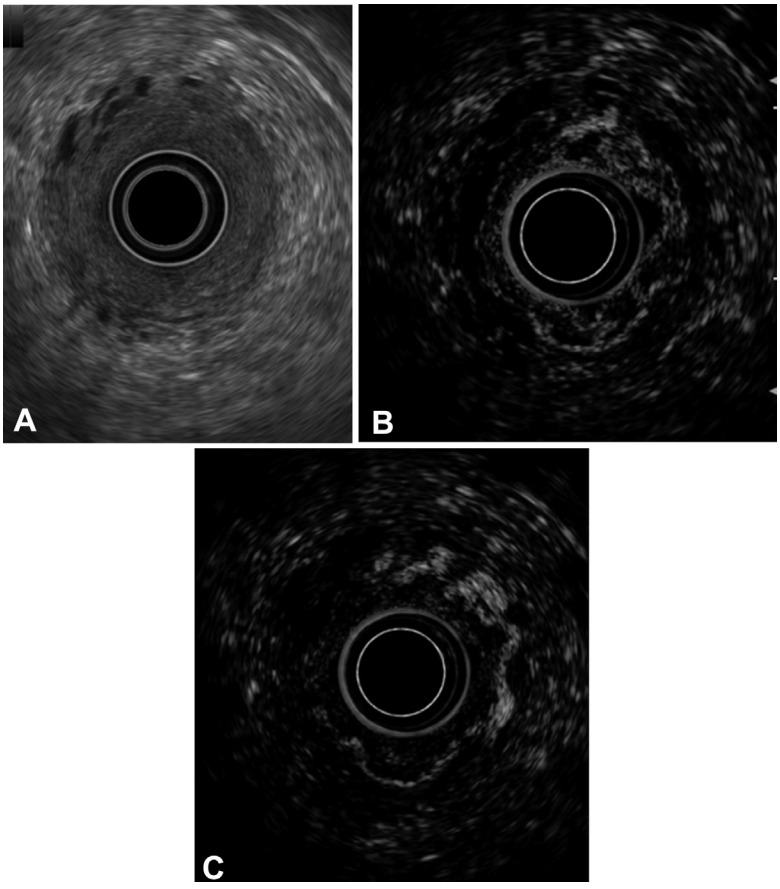

Figure 4. EUS revealed rectal wall hyperechoic thickening with unclear layer structures. **A,** Small anechoic areas were observed in and around the rectal wall. **B,** Contrast-enhanced EUS revealed a septum-like structure and **(C)** subsequent slow flow into the cavity of the thickened wall. Anechoic areas were not enhanced.

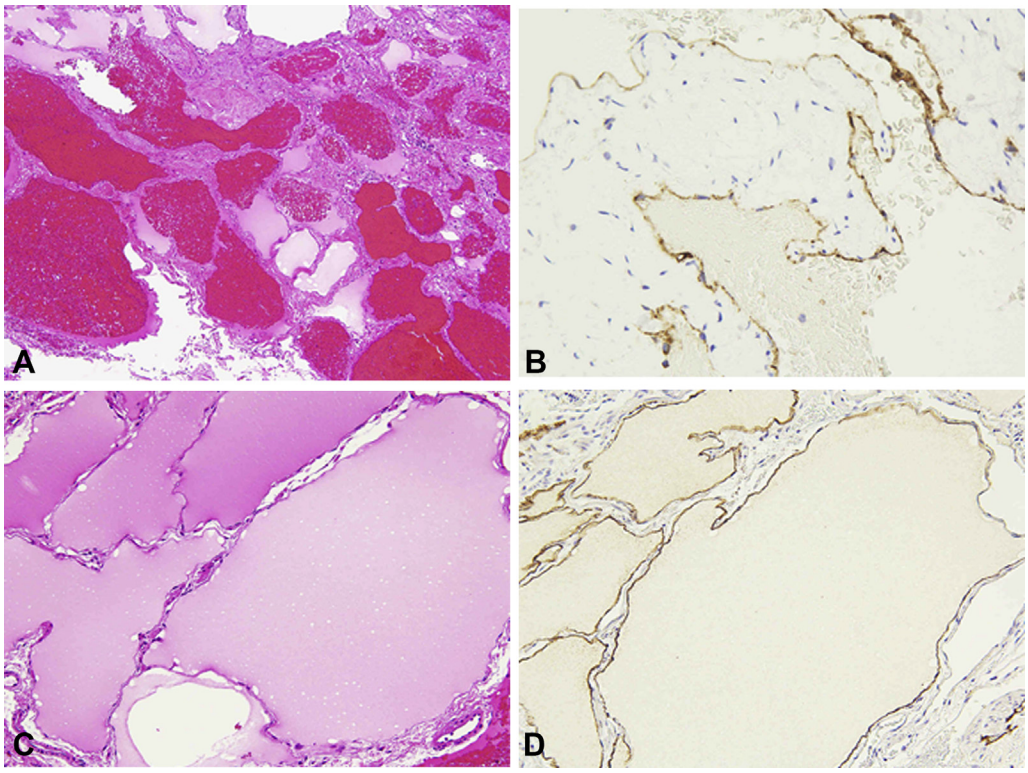

Figure 5. Histologic findings of surgical specimen. **A, B,** The irregularly dilated vascular overgrowth consisted of 2 components. **C, D,** Hemangioma contained red blood cells and was lined by CD31-positive cells. Lymphangioma contained eosinophilic liquid lined by D2-40–stained endothelial cells.

rarity, awareness of DCH as a cause of hematochezia is important for early diagnosis.

As a side note, recent reports demonstrated that elastosonography also aids in the noninvasive differentiation of hemangioma and other lesions in the liver and skin.[6,7] The efficacy of EUS modalities, including contrast-enhanced EUS and elastosonography, in the diagnosis of intestinal submucosal lesions, such as DCH, needs to be further evaluated.

DISCLOSURE

All authors disclosed no financial relationships.

Abbreviation: DCH, diffuse cavernous hemangioma.

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