

Local rectal portal hypertension in the absence of a patent superior rectal vein

A 16-year-old girl was referred to our unit for recurrent anemia caused by rectal bleeding that had required regular blood transfusions since her birth. The patient's history did not reveal any other abnormalities such as inflammatory disease or perinatal complications. Physical examination revealed the presence of grade III hemorrhoids and abnormal enlargement of the clitoris (● Fig. 1). Colonoscopy showed diffuse submucosal venous dilatation in the perianal region, rectum, and low sigmoid colon (● Fig. 2). Magnetic resonance imaging (MRI) with angiography demonstrated dilatation mainly of the venous mesorectal network and multiple opacifications in the soft tissues that showed uptake of contrast agent before there was uptake in the systemic venous system (● Fig. 3). The iliac veins showed a normal blood flow, while the patency of the superior rectal vein could not be clearly distinguished.

The features of this patient's clinical presentation are suggestive of a rectal portal cavernoma on the basis of local rectal portal hypertension. Embolization was discussed but not proposed because of the potential absence of venous drainage through the inferior mesenteric vein. The patient underwent a laparoscopic exploration, which revealed the absence of a patent superior rectal vein, this being the main branch of the inferior mesenteric vein (● Video 1). A laparoscopic low anterior resection with a coloanal anastomosis and temporary diverting loop ileostomy was performed, the only complication being a pelvic hematoma that

spontaneously drained through the vagina and anus, which delayed definitive closure of the protective ileostomy.

The ileostomy was closed after 6 months. Since that time, encompassing a follow-up of 1 year since closure of the ileostomy, the patient has not experienced any transanal bleeding and has had no problems with fecal continence. There is no soiling and she passes stools three to six times a day without fragmentation and emergencies. The clitoris, which was hypertrophied, has decreased in size by more than half.

Different types of congenital portal venous malformations have been reported in the literature [1]. Generalized portal hypertension has been described as causing hemorrhoids in the newborn [2]. Despite reports of the absence of an inferior mesenteric vein [3], this is the first case suggestive of the absence of a patent superior rectal vein as the underlying etiology (● Fig. 4). Potential causes include chronic thrombosis of the superior rectal vein leading to an obliterated vein and an atrophic tract. Thrombosis can result secondarily from a prothrombotic state, thrombophilia, or a local intra-abdominal infection causing obliteration of the vein and subsequently an atrophic tract [4,5]. Neonatal portal vein thrombosis has been reported in the setting of umbilical venous catheterization or peripartum asphyxia [5].

The above-mentioned etiology must be distinguished from other types of more common vascular malformations such as vascular ectasia or hemangioma, because

Video 1



Views during laparoscopic exploration and low anterior resection with a coloanal anastomosis showing local rectal portal hypertension at the level of the distal sigmoid, rectum, and upper part of the anal canal. The left colic vein drains into the inferior mesenteric vein without the presence of a patent superior rectal vein. Instead a dilated venous mesorectal network can be seen. The superior rectal artery is divided.

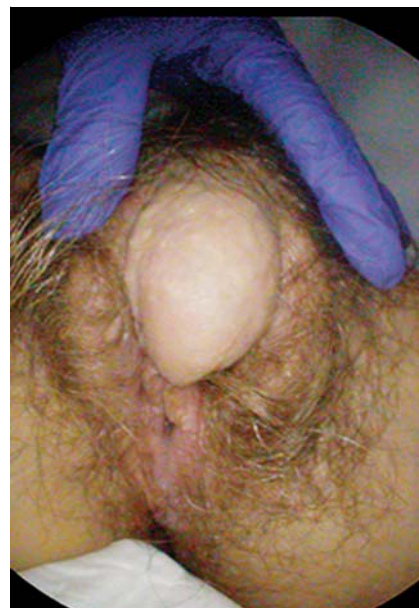


Fig. 1 Abnormal enlargement of the clitoris in a 16-year-old girl who had recurrent rectal bleeding and hemorrhoids.

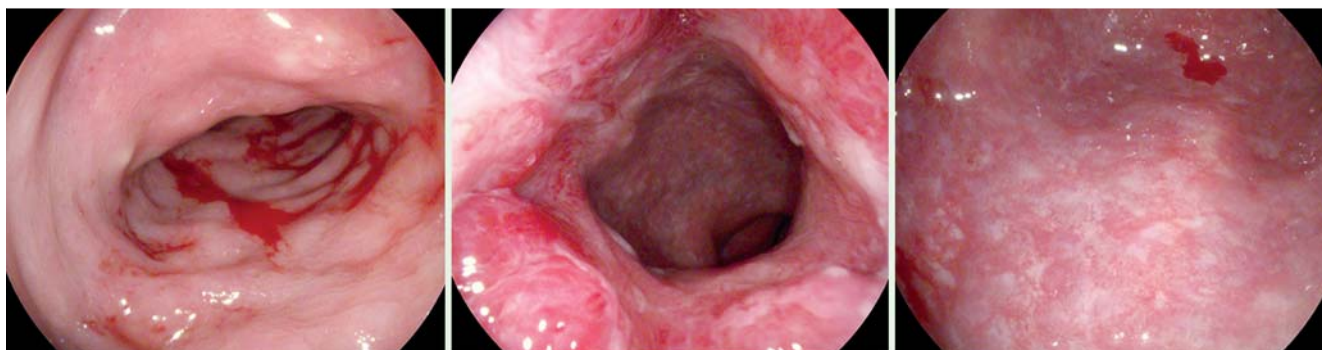


Fig. 2 Colonoscopic views showing diffuse submucosal venous dilatation in the perianal region, rectum, and low sigmoid colon.



Fig. 3 Magnetic resonance angiography demonstrates dilatation mainly of the venous mesorectal network and multiple opacifications in the soft tissues, which show contrast agent uptake before uptake in the systemic venous system: **a** in sagittal view; **b** in coronal view.

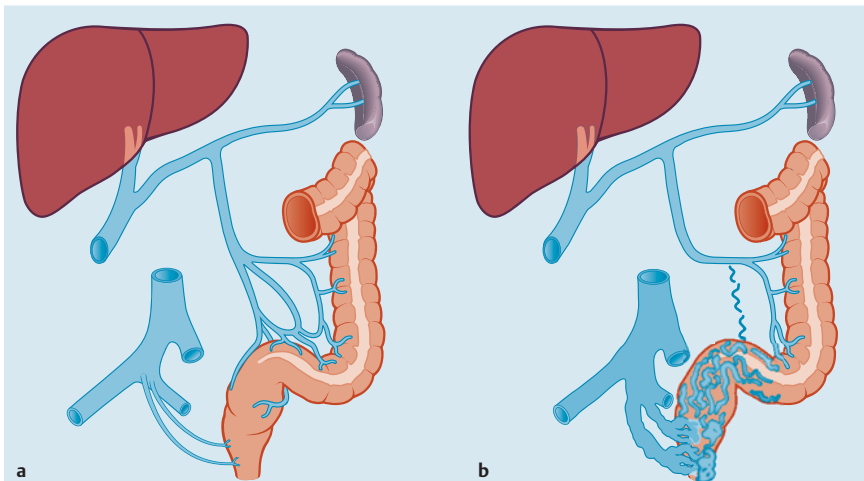


Fig. 4 Schematic drawing of the relevant intra-abdominal vasculature. **a** The classical anatomy as depicted by anatomical textbooks; **b** the anatomy in the current case with absence of a patent superior mesenteric vein, which resulted in local sigmoid and rectal portal hypertension.

treatment modalities for this type of disease, such as embolization, will cause venous rectal ischemia. The likelihood of this type of venous malformation might

be higher than suspected in the literature and needs to be recognized.

Endoscopy_UCTN_Code_CCL_1AD_2AI

Competing interests: None

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DOI <http://dx.doi.org/10.1055/s-0042-108571>
 Endoscopy 2016; 48: E226–E227
 © Georg Thieme Verlag KG
 Stuttgart · New York
 ISSN 0013-726X

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