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 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.

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

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.
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.

Competing interests: None

Joel Leroy1, Arthur R. Wijsmuller2, Edris Wedi2, Catherine Roy3, Didier Mutter1, Juergen Hochberger2

1 IRCAD/ EITS, Department of General, Digestive and Endocrine Surgery, Nouvel Hôpital Civil, University Hospital of Strasbourg, Strasbourg, France
2 Department of Gastroenterology, Nouvel Hôpital Civil, University Hospital of Strasbourg, Strasbourg, France
3 Department of Radiology, Nouvel Hôpital Civil, University Hospital of Strasbourg, Strasbourg, France

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Corresponding author
Joel Leroy, MD
Department of Digestive and Endocrine Surgery Nouvel Hôpital Civil PO Box 67000 Strasbourg France
joel.leroy@ircad.fr

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 rectal vein, which resulted in local sigmoid and rectal portal hypertension.