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Klippel-Trénaunay Syndrome with Involvement of Coecum and Rectum: A Rare Cause of Lower Gastrointestinal Bleeding

T. Mussack[1], J. T. Siveke[2], K. J. Pfeifer[3], C. Folwaczny[1,2]

[1]Department of Surgery Innenstadt, [2]Medizinische Klinik Innenstadt,
[3]Institut für Klinische Radiologie, Klinikum der Universität München, Germany

Abstract: Klippel-Trénaunay syndrome (KTS) is a congenital malformation usually presenting limb asymmetry, abnormal development of the deep and superficial veins, and cutaneous capillary malformations. We describe the case of a 56-year-old male KTS patient who suffered from recurrent but life non-threatening lower gastrointestinal bleeding. Colonoscopy revealed multiple extensive cavernous hemangiomas in the coecum and the ascending colon as well as the sigmoid colon and the rectum. MR imaging showed numerous dilated vessels within the left gluteal and inguinal region. The mucosal and the submucosal layers particularly of the sigmoid colon and rectum appeared markedly broadened and displayed high signal intensities in the STIR sequences. Due to only moderate oozing at time of admission the patient was treated with oral iron supplementation so far.

Key words: Klippel-Trénaunay syndrome; hemangioma; gastrointestinal bleeding

Introduction

Klippel-Trénaunay syndrome (KTS) is rare congenital malformation characterized by a) bony or soft tissue hypertrophy, usually affecting one extremity, b) hemangiomas and / or lymphangiomas and c) varicosis or venous malformations [6, 11]. Vascular malformations of the lower GI tract in KTS have been reported only in single cases [7, 11]. The clinical manifestations range from occult to massive, life-threatening hemorrhages, which can be usually treated only by resection of the involved bowel segment [10]. The situation is complicated when other pelvic structures are involved, especially in young adults. Endoscopic therapy has a limited role because of the commonly diffuse nature of the intestinal hemangiomas [1, 8]. We present this rare case of diffuse hemangiomas mainly of the coecum and the rectum as a cause of recurrent lower GI bleeding in a KTS patient.

Case Report

A 56-year-old male presented with a 3-month history of occasional lower gastrointestinal bleeding following defecation. The diagnosis of a Klippel-Trénaunay syndrome had been already established during childhood. Furthermore, the patient reported on recurrent self-limiting bleeding caused by gentle palpation of cutaneous hemangiomas located at the left lateral thigh and the perineum. Thirteen years before admission he underwent emergency surgery due to a semicircular left-sided perineal abscess with an intersphincteric fistula. This operation was reported to take more than 3 hours due to uncontrollable bleeding. Two years later, acute gross hematuria due to bladder hemangiomas was successfully treated by laser coagulation.

At the time of admission laboratory evaluation was compatible with mild iron deficient anemia (hemoglobin 10g/dl), whereas the platelet count and coagulation parameters were normal. General physical and abdominal examinations were unremarkable. Colonoscopy demonstrated subtotal circular covering of the colonic wall by extensive cavernous angiomas, which were mainly found in the coecum and the ascending colon (Fig. 1) as well as the sigmoid colon and the rectum and spared the descending colon and the terminal ileum.

Subsequent esophagogastroduodenoscopy revealed no pathologic findings. MR imaging of the pelvic region showed numerous dilated vessels within the left

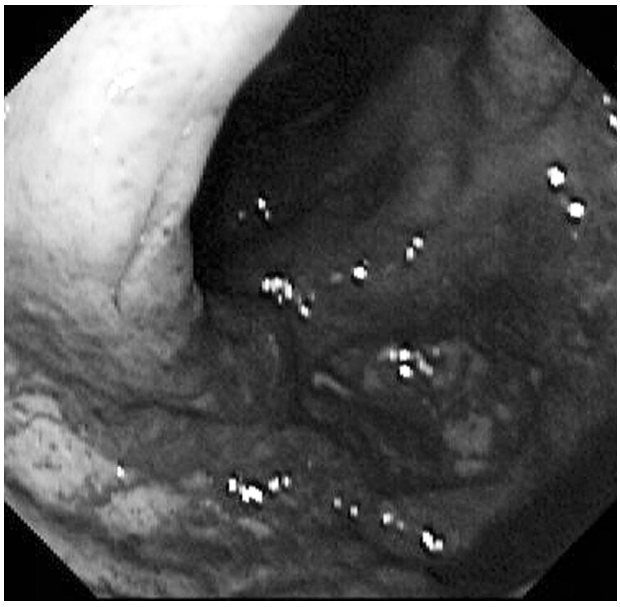

Fig. 1. Colonoscopy revealing extensive cavernous angiomatosis of the coecum, the ascending and the transverse colon.

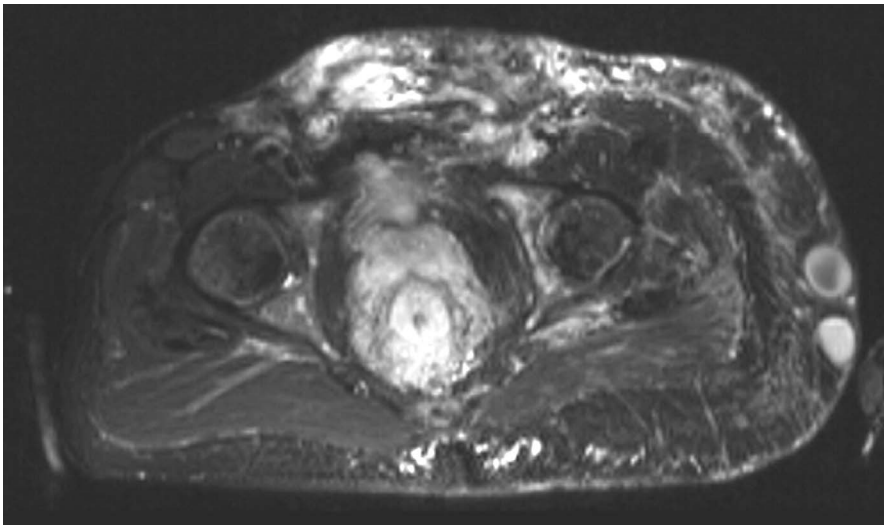

Fig. 2. MR imaging using STIR sequences showing diffuse angiomatous dilation of rectal vessels and thickening of the mucosal and the submucosal layers of the rectal wall.

gluteal and inguinal region with a maximum diameter of 35 mm. The mucosal and the submucosal layers of the sigmoid colon and rectum appeared markedly broadened and displayed high signal intensities in the STIR sequences (Fig. 2). In addition, angiomas were also found in the prostate gland.

Selective digital subtraction angiography of the superior and inferior mesenteric arteries showed multiple dilated vessels with contrast-enhanced lacunas particularly along the ascending as well as the sigmoid colon and the rectum.

Dicussion

In one of the largest published series of KTS patients, hematochezia was reported in only six of 588 patients, although a few other cases may have gone unnoticed [10]. The most common cause of GI bleeding is attributed to diffuse cavernous hemangiomas of the distal colon and rectum, found in estimated 1 – 12.5% of KTS patients. Although quite rare, bleeding from jejunal hemangiomas have also been described [2, 9]. Usually, GI hemorrhage in KTS patients begins in the first decade of life and tends to be intermittent in nature [4, 10].

The pathogenesis of visceral hemangiomatosis is still unclear. The interruption of the superficial femoral or popliteal veins of the affected limb possibly results in shunting of the blood flow into dilated internal iliac veins, hampering drainage of vesical, genital and rectal veins with subsequent venous malformations in their respective organs [10]. However, this hypothesis does not explain the presence of vascular malformations found in the small intestine or the portal system [2]. Therefore, KTS is supposed to be more likely a manifestation of a generalized mesodermal development abnormality [11].

The role of MR angiography has not been well defined in KTS but has the potential of assessing these vascular malformations with better accuracy [5]. Nevertheless, a visceral angiogramm is still required particularly preoperatively for defining the anatomy and the extent of the intestinal involvement to guide surgical resection [3].

Endoscopic examination may reveal visible mucosal vessels or compressed nodules as well as extensive bluish angiomatous submucosal lesions. However, the extent of involvement can be only limitedly determined by visualization of the mucosal anomalies. Lesions in deeper layers of the intestinal wall cannot be well assessed. Biopsies of suspected hemangiomas should be avoided, as it may precipitate severe hemorrhages [11].

Management of colorectal intestinal hemangiomas in KTS will depend on the extent and severity of blood loss. When the entire rectum is severely involved, surgery is less attractive, as a permanent stoma will prove necessary. Conservative management and iron supplements may be sufficient in those patients who present with occasional nonsignificant and nondebilitating bleedings. However, long-term treatment mostly requires surgical resection comprising proctocolectomy in the case of life-threatening colonic bleeding and coloanal anastomosis with preservation of anal function, especially in younger patients, or abdominoperineal resection in the case of rectal bleeding due to the diffuse and sometimes progressive disease process [11]. Vascular embolization can be considered if a distinct bleeding site is encountered. Endoscopic photocoagulation using argon laser is sometimes employed for the management of localized lesions or ablation of postoperative residual disease. The latter has provided effective palliation of colorectal venous angiomas with minimal morbidity [1, 8].

In summary, this report describes extensive cavernous hemangiomas of the entire colon excluding the distal transverse and descending colon in a patient with Klippel-Trénaunay syndrome as a rare cause of recurrent lower gastrointestinal bleeding. Apart from occasional hematochezia and mild anemia our patient was asymptomatic. Furthermore, with respect to the extensive colonic involvement described herein bleeding will most likely only subside following surgical resection of the entire colon. Hence, only oral iron supplementation was administered so far.

REFERENCES

1. Azizkhan RG (1991) Life-threatening hematochezia from a rectosigmoid vascular malformation in Klippel-Trenaunay syndrome: long-term palliation using an argon laser. J Pediatr Surg 26: 1125-1127
2. Brown R, Ohri SK, Ghosh P, Jackson J, Spencer J, Allison D (1991) Case report: jejunal vascular malformation in Klippel-Trenaunay syndrome. Clin Radiol 44: 134-136
3. Gandolfi L, Rossi A, Stasi G, Tonti R (1987) The Klippel-Trenaunay syndrome with colonic hemangioma. Gastrointest Endosc 33: 442-445
4. Ghahremani GG, Kangarloo H, Volberg F, Meyers MA (1976) Diffuse cavernous hemangioma of the colon in the Klippel-Trenaunay syndrome. Radiology 118: 673-678
5. Haber M, Reuben A, Burrell M, Oliverio P, Salem RR, West AB (1995) Multiple focal nodular hyperplasia of the liver associated with hemihypertrophy and vascular malformations. Gastroenterology 108: 1256-1262
6. Klippel M. Trénaunay P (1900) Du noevus variqueux ostéohypertrophique. Arch Gen Med 185: 641-672
7. Lehmann TG, Dux M, von Herbay A, Lehnert T (2000) [Klippel-Trenaunay syndrome with involvement of the rectum. Surgical therapy after interventional-radiologic preparation]. Chirurg 71: 228-233
8. Myers BM (1993) Treatment of colonic bleeding in Klippel-Trenaunay syndrome with combined partial colectomy and endoscopic laser. Dig Dis Sci 38: 1351-1353
9. Schmitt B, Posselt HG, Waag KL, Muller H, Bender SW (1986) Severe hemorrhage from intestinal hemangiomatosis in Klippel-Trenaunay syndrome: pitfalls in diagnosis and management. J Pediatr Gastroenterol Nutr 5: 155-158
10. Servelle M, Bastin R, Loygue J, Montagnani A, Bacour F, Soulie J, Andrieux JB (1976) Hematuria and rectal bleeding in the child with Klippel and Trenaunay syndrome. Ann Surg 183: 418-428
11. Wilson CL, Song LM, Chua H, Ferrara M, Devine RM, Dozois RR, Nehra V (2001) Bleeding from cavernous angiomatosis of the rectum in Klippel-Trenaunay syndrome: report of three cases and literature review. Am J Gastroenterol 96: 2783-2788

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Address for correspondence:
Thomas Mussack, M.D.
Department of Surgery Innenstadt
Klinikum der Universität München
Nussbaumstrasse 20
D-80336 München, Germany
Tel: +49-89-5160-2638
Fax: +49-89-5160-4489
E-mail: Thomas.Mussack@med.uni-muenchen.de