Turk J Gastroenterol 2006; 17 (4): 308-312

Diffuse cavernous hemangioma of the rectosigmoid colon with extraintestinal involvement

Ekstraintestinal tutulum gösteren rektosigmoid kolon diffüz kavernöz hemanjiomu

Ömer TOPALAK[1], Can GÖNEN[1], Funda OBUZ[2], Mustafa SEÇİL[2]

Departments of [1]Gastroenterology, [2]Radiology, Dokuz Eylül University School of Medicine, Izmir

Cavernous hemangioma of the colon is a rare cause of gastrointestinal bleeding. These lesions can be encountered as solitary, multiple, or part of a more complex syndrome with cutaneous manifestations. We herein describe a 26-year-old woman with cavernous hemangioma involving the rectosigmoid area. Additional hemangiomas were identified in the pelvic structures, spine, iliac bone and spleen. This multi-visceral involvement without cutaneous manifestations represents an intermediate variety between solitary hemangioma and well-defined syndromes with cutaneous and structural anomalies. The potential presence of extraintestinal hemangiomatosis should be considered and investigated in patients with cavernous hemangioma of the colon even without cutaneous manifestations or with a limited colonic involvement.

Key words: Cavernous hemangioma, extraintestinal involvement, hemangiomatosis

Kolonun kavernöz hemanjiomu nadir bir gastrointestinal kanama nedenidir. Bu lezyonlar soliter, çok sayıda veya kutanöz bulgularla birliktelik gösteren kompleks bir sendromun parçası olabilirler. Bu yazıda, rektosigmoid bölgede kavernöz hemanjiom saptanan 26 yaşında bir bayan hasta sunulmuştur. Pelvik yapılar, omurga, iliak kemik ve dalakta eşlik eden hemanjiomlar saptanmıştır. Kutanöz bulgular olmadan bu şekildeki bir multi-viseral tutulum, kutanöz ve yapısal anomaliler ile birliktelik gösteren iyi tanımlanmış sendromlar ve soliter lezyonlar arasında intermedier bir formu tanımlamaktadır. Kolonun kavernöz hemanjiomu olan hastalarda, kolonda sınırlı bir tutulum olması veya kutanöz bulgular saptanmamasına rağmen ekstraintestinal hemanjiomatozis olabileceği akılda tutulmalıdır.

Anahtar kelimeler: Kavernöz hemanjiom, ekstraintestinal tutulum, hemanjiomatozis

INTRODUCTION

Vascular lesions of the gastrointestinal tract are being recognized with increasing frequency as a cause of gastrointestinal bleeding. Cavernous hemangioma of the colon is a rare entity in this spectrum, grouped with benign vascular malformations of the gastrointestinal tract (1). Both solitary and multiple hemangiomas within the gastrointestinal tract and colonic hemangioma-related syndromes with well-defined cutaneous manifestations, such as Klippel-Trenaunay-Weber or blue rubber bleb nevus syndrome, have been described in the literature (2-4). We herein describe a patient with cavernous hemangioma of the rectosigmoid area with extraintestinal hemangiomatosis affecting pelvic structures, spine and spleen, representing an intermediate variety in this spectrum. A brief description of diagnostic characteristics and imaging modalities, emphasizing the extraintestinal involvement, is given in this report.

CASE REPORT

A 26-year-old woman with a 10-year history of recurrent and intermittent rectal bleeding presented to our gastroenterology clinic. The bleeding coated the outside of the stool and was self-limited in nature. The patient had no additional symptoms. Six years before the current admission, diagnosis of ulcerative colitis and iron deficiency anemia had been made at a local hospital and a four-month treatment trial with oral mesalamine resulted in no improvement. She had been on oral and parenteral iron supplementation therapy since then. Physical examination revealed a pale woman with conjunctival pallor and no evidence of

Address for correspondence: Ömer TOPALAK
Department of Gastroenterology, Dokuz Eylül University School of Medicine, 35340, İnciraltı, Izmir, Turkey
Phone: +90 232 412 37 06 • Fax: +90 232 259 05 41
E-mail: omer.topalak@deu.edu.tr

Manuscript received: 04.04.2006 **Accepted:** 18.09.2006

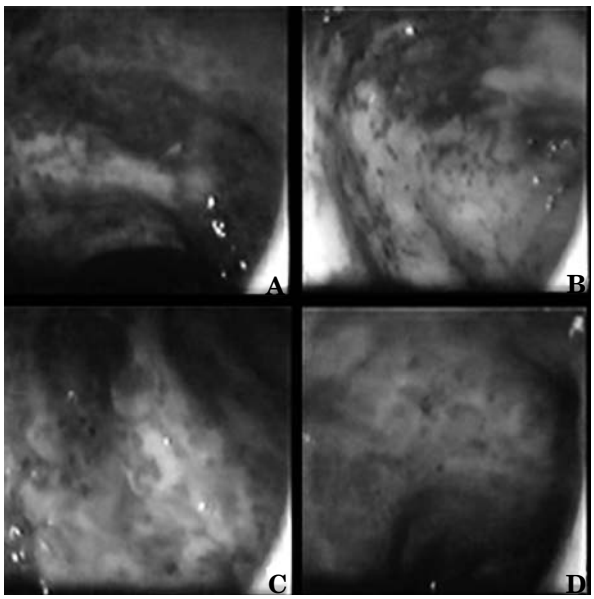

Figure 1. A-D) Colonoscopic image of the cavernous hemangioma, proximal border (A-C) and central part (D)

mucocutaneous vascular lesions. The rest of the examination was unremarkable. Initial laboratory tests yielded the following results: hemoglobin 10.4 g/dl, mean corpuscular volume 72.4 fL, platelet count 250,000/µL, transferrin saturation 6.7%, ferritin 4 µg/L, and C-reactive protein 0.28 mg/dl.

Colonoscopy revealed a dark purple-bluish and dull red, raised, contiguous lesion extending from the anal canal to the sigmoid colon (Figure 1). There were erosions with slight oozing. Biopsy specimens were not obtained because of the risk of inducing bleeding. Furthermore, the appearance was quite characteristic of diffuse cavernous hemangioma. Multislice computed tomography of the pelvis disclosed marked thickening of the rectosigmoid wall with multiple phleboliths. The lesion was hypovascular during arterial phase and there were enlarged venous structures in the pelvic region. Similar thickening was noticed at the uterine cervix (Figure 2). Magnetic resonance imaging (MRI) showed diffuse circumferential rectal wall thickening with high signal intensity on T2-weighted images. Although uterine cervix, distal part of the fundus and vagina appeared enlarged and homogeneously hyperintense (Figure 3), these involvements were not considered to be direct infiltration of the rectal lesion. Additional hemangiomas were identified in the first sacral vertebra and in the right iliac bone. Multiple hemangiomas,

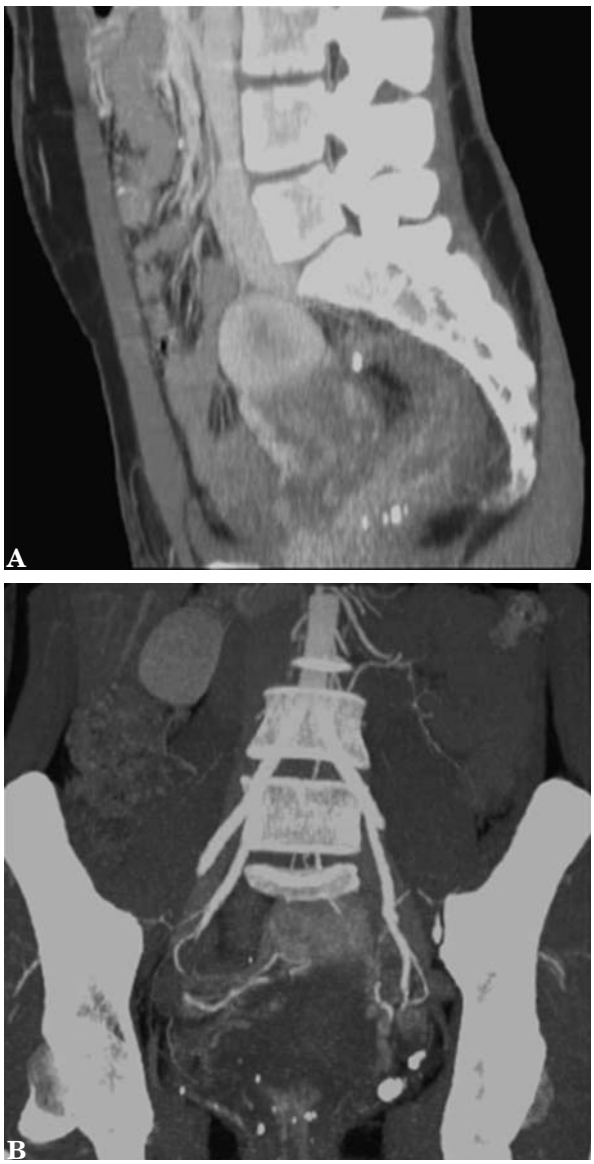

Figure 2. A-B) Sagittal maximum intensity projection (MIP) CT angiographic image obtained during arterial phase reveals diffuse rectal wall thickening and uterine cervix enlargement. There are multiple phleboliths in the rectum and uterine cervix (A). Coronal MIP CT angiographic image obtained during arterial phase showing the hypovascular lesions (B)

with the largest 2 cm in diameter, were also present in the spleen (Figure 4). A diagnosis of cavernous hemangioma of the rectosigmoid area with extraintestinal hemangiomatosis was made. The patient refused gynecologic examination, and elected to receive conservative follow-up rather than to undergo curative surgical intervention. When last seen at her 12-months follow-up, there had been no major hemorrhagic episode.

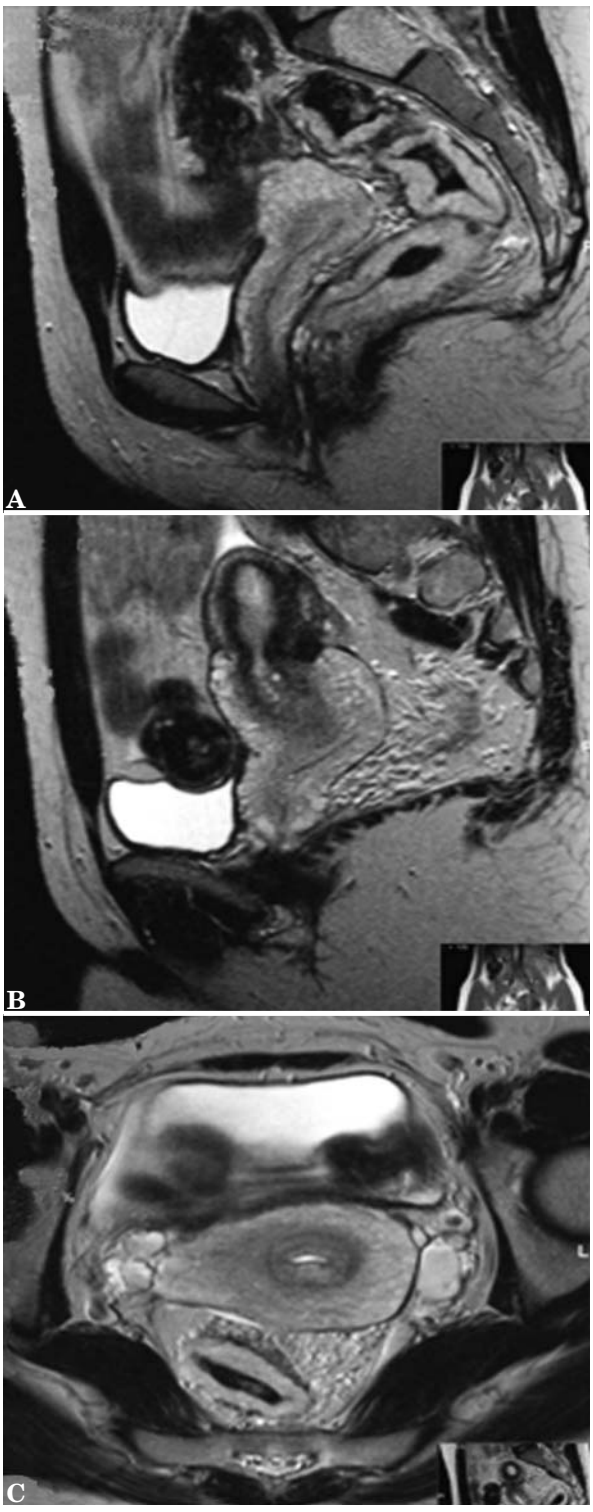

Figure 3. A-C) Turbo spin echo T2-weighted sagittal MR images demonstrating diffuse concentric wall thickening of the rectosigmoid colon. Uterine cervix and vagina are enlarged, but uterine corpus is intact. The lesions are homogeneously hyperintense (A-B). Turbo spin echo T2-weighted transverse MR image showing rectal wall thickening and uterine cervix enlargement (C)

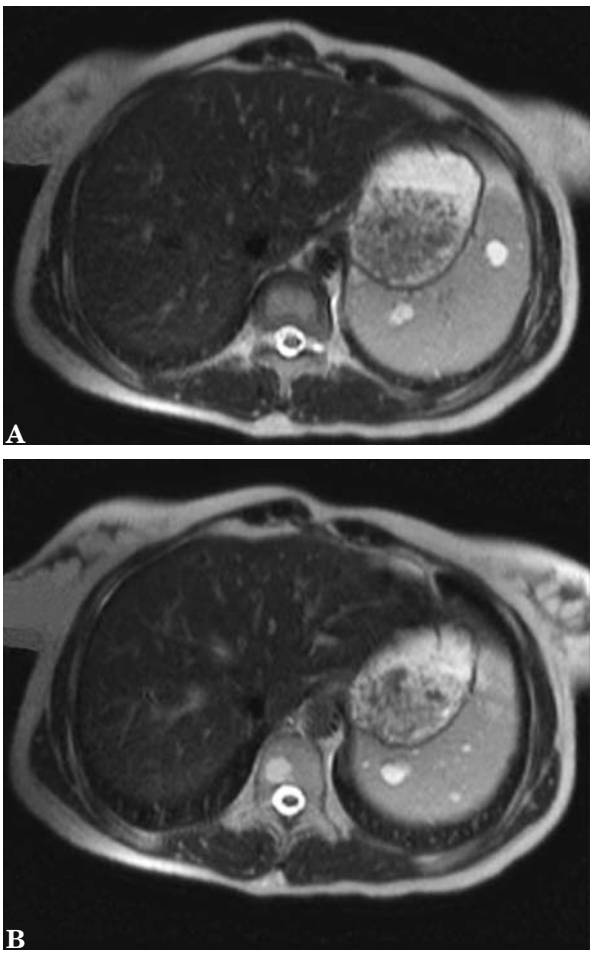

Figure 4. A-B) Turbo spin echo single shot T2-weighted transverse MR images demonstrating splenic (A) and vertebral hyperintense hemangiomas (B)

DISCUSSION

Cavernous hemangioma of the rectosigmoid colon is a rare benign vascular lesion (1). A survey of 9000 colonoscopies performed in our institution has revealed only two cases during a study period of 13 years. Histologically, cavernous hemangiomas are composed of numerous dilated, thin-walled, irregular blood-filled spaces mainly located within the mucosa and submucosa, sometimes extending through the muscular layer to the serosa (4). These lesions can be classified as hamartomas between malformations and tumors (1). Recurrent, painless rectal bleeding is the most common clinical presentation and more than half of the patients have some degree of iron deficiency anemia (2). A substantial number of patients are misdiagnosed and undergo unnecessary surgical procedu-

res before an accurate diagnosis is made (5). Likewise, a misdiagnosis of ulcerative colitis was made in our patient. Internal hemorrhoids, adenomatous polyps and carcinoma constitute other mimicking lesions that have been reported (6). Moreover, correct diagnosis delays can be due to multiple morphological characteristics of the lesions and to lack of knowledge. Öner and Altaca (7) reported an average delay of 19 years between the appearance of the initial symptoms and diagnosis. Colonoscopy is essential for the diagnosis, and elevated dark purple-red nodules or dilated, engorged vessels are characteristic findings (Figure 1). The rectosigmoid area is the most common site of cavernous hemangioma in the gastrointestinal tract (2). Moreover, diffuse colonic hemangiomatosis has also been described (3). Massive rectal bleeding, Kasabach-Merritt syndrome, pelvic organ infiltration (e.g. bladder and uterus) causing hydroureteronephrosis or vessel compression, and fistula formation have been reported as complications (5, 8-10). In this context, imaging has an important role for both diagnostic and therapeutic purposes. The presence of phleboliths on abdominal plain films and a narrowed, scalloping rigid rectal lumen with widened presacral space on barium enema could suggest the diagnosis (4). Computed tomography scans can give a more accurate diagnosis by demonstrating thickened rectosigmoid wall, phleboliths and pelvic invasion. By clearly delineating the extent, invasion and extraintestinal involvement of cavernous hemangiomas, MRI has become the diagnostic procedure of choice for preoperative evaluation (2). Characteristically, cavernous hemangioma demonstrates bright signal intensity on T2-weighted images and intermediate signal intensity on T1-weighted images (Figure 3). Calcifications and blood vessels are signal voided on both T1- and T2-weighted images, but thrombosed vessels have high signal intensity. Moreover, endorectal surface coil MRI and endosonography depicts bowel wall and sphincter muscles more clearly. This could be useful for sphincter-saving surgical procedures (11).

The only effective treatment for cavernous hemangioma is complete resection of the involved colonic segment. If technically feasible, a sphincter-saving procedure with anterior resection and coloanal anastomosis is most appropriate. Abdominoperineal resection is performed when the hemangioma extends into the anal canal where sphincters are involved (1, 5, 7). Nonoperative techniques such as sclerotherapy, electrocautery, cryosurgery and radium implantation have been used in selected cases and usually provide only temporary relief. Angiography and embolization should be used only in cases of acute bleeding; the technique is generally regarded as unreliable as bleeding recurs in most patients (2, 10, 11).

Multiple hemangiomas of the skin, generally encountered in infancy, have traditionally been recognized as a clue to visceral hemangiomas. Subcategories according to cutaneous involvement, such as "segmental" or "disseminated neonatal" types, have a high associated risk of visceral involvement, although many of these patients also have some other structural anomalies (e.g. brain malformations, arterial anomalies and cardiac defects). Also, the liver, not the gastrointestinal tract, is the most common site of visceral involvement in these cases (12, 13). Furthermore, colonic hemangiomas can be encountered in two well-defined syndromes. In blue rubber bleb nevus syndrome, cutaneous blue-colored, wrinkled, easily compressible vascular lesions are related to intestinal hemangiomas. In Klippel-Trenaunay-Weber syndrome, cutaneous hemangioma, hypertrophy of the bones and soft tissues of the involved limb and varicose veins limited to the affected side could be accompanied by colonic hemangiomas (4). The patient presented here - with extraintestinal involvement and with no cutaneous manifestations or any syndrome-related findings – can be considered on the border between well-defined visceral hemangiomatosis-related syndromes and isolated cavernous hemangioma of the colon.

In conclusion, cavernous hemangioma of the rectosigmoid area is an uncommon vascular lesion typically presented as painless rectal bleeding with characteristic endoscopic findings. The potential presence of extracolonic hemangiomatosis should be considered in patients with cavernous hemangioma of the colon even without cutaneous manifestations or with a limited colonic involvement.

REFERENCES

1. Pohlen U, Kroesen AJ, Berger G, Buhr HJ. Diagnostics and surgical treatment strategy for rectal cavernous hemangiomas based on three case examples. Int J Colorectal Dis 1999; 14: 300-3.
2. Yorozuya K, Watanabe M, Hasegawa H, et al. Diffuse cavernous hemangioma of the rectum: report of a case. Surg Today 2003; 33: 309-11.

3. Dobozi BM, Rockey DC. Diffuse colonic hemangiomatosis. Gastrointest Endosc 2004; 60: 799.
4. Greenwald DA, Brandt LJ. Vascular lesions of the gastrointestinal tract. In: Feldman M, Friedman LS, Sleisenger MH, eds. Sleisenger & Fordtran's Gastrointestinal and Liver Disease. Pathophysiology/Diagnosis/Management. 7th ed. Philadelphia: Saunders, 2002; 2341-55.
5. Tan TC, Wang JY, Cheung YC, Wan WY. Diffuse cavernous hemangioma of the rectum complicated by invasion of pelvic structures. Report of two cases. Dis Colon Rectum 1998; 41: 1062-6.
6. Hsu RM, Horton KM, Fishman EK. Diffuse cavernous hemangiomatosis of the colon: findings on three-dimensional CT colonography. Am J Roentgenol 2002; 179: 1042-4.
7. Oner Z, Altaca G. Diffuse cavernous rectal hemangioma--clinical appearance, diagnostic modalities and sphincter saving approach to therapy: report of 2 and a collective review of 79 cases. Acta Chir Belg 1993; 93: 173-6.
8. Aylward CA, Orangio GR, Lucas GW, Fazio VW. Diffuse cavernous hemangioma of the rectosigmoid--CT scan, a new diagnostic modality, and surgical management using sphincter-saving procedures. Report of three cases. Dis Colon Rectum 1988; 31: 797-802.
9. Pierie JP, van Vroonhoven TJ. Cavernous hemangioma of the rectosigmoid. J Am Coll Surg 2000; 190: 750.
10. Lyon DT, Mantia AG. Large-bowel hemangiomas. Dis Colon Rectum 1984; 27: 404-14.
11. Hasegawa H, Teramoto T, Watanabe M, et al. Diffuse cavernous hemangioma of the rectum: MR imaging with endorectal surface coil and sphincter-saving surgery. J Gastroenterol 1996; 31: 875-9.
12. Metry DW, Hawrot A, Altman C, Frieden IJ. Association of solitary, segmental hemangiomas of the skin with visceral hemangiomatosis. Arch Dermatol 2004; 140: 591-6.
13. Holden KR, Alexander F. Diffuse neonatal hemangiomatosis. Pediatrics 1970; 46: 411-21.