DIFFUSE CAVERNOUS HEMANGIOMA OF THE RECTUM (DCHR) – DIAGNOSIS AND TREATMENT – CASE REPORT AND REVIEW OF AVAILABLE LITERATURE

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Diffuse cavernous hemangioma of the rectum (DCHR) is a rare benign tumor of vascular origin. Approximately 200 such cases have been reported in the literature. Here we present a case of a 49-year old female patient who underwent a surgical procedure due to a mass of the rectum with a history of recurrent, painless gastrointestinal bleeding and anemia in whom DCHR was diagnosed postoperatively. This paper is intended as a metaanalysis of available diagnostic and therapeutic methods to be used in patients with DCHR.

Key words: DCHR, cavernous hemangioma of the rectum

Diffuse cavernous hemangioma of the rectum (DCHR) is a rare benign tumor of vascular origin (1, 2, 4, 6). Pathogenesis of this disorder has not been fully elucidated (3). Formation of DCHR is suspected to stem from abnormal development of mosodermal tissue in the prenatal period (3, 4). DCHR was reported for the first time by Philip in 1839 (3, 6). So far approximately 200 such cases have been reported in the literature. Cavernous hemangiomas account for approximately 80% of hemangiomas of the rectosigmoid region (1). From the morphological point of view, these are massive cavities lined with one or multiple epithelial layers (1). Most commonly DCHR is diagnosed in patients between 5 and 25 years of age (3) with a history of multiple, painless, rectal bleeding episodes or an episode of massive, life threatening gastrointestinal bleeding and anemia (1, 2, 3, 4). Due to nonspecific symptoms and complex diagnostic workup, delay in the diagnosis of DCHR amounts to approximately 19 years in this patient group (6, 7). This paper is intended as an analysis of available diagnostic and therapeutic methods to be used in patients with DCHR.

CASE REPORT

A 49-year old female patient, in good general condition, was admitted to the department to undergo an elective surgical treatment of a rectal mass. The mass did not cause any clinical symptoms, was found in CT imaging 2 months before admission to the department. CT imaging demonstrated a pathological tissue mass, 15 x 10 x 9.5 cm in size, located in the rectum, reaching the anal sphincter, with enlarged local lymph nodes. Digital rectal ex-
amination demonstrated a palpable, compliant mass, that looked like reaching the sphincters. Bilaterally enlarged, painful inguinal lymph nodes. Colonoscopy was not done. Tumor markers were normal. She had a history of several gastrointestinal bleeding episodes, recurrent anemia (at admission her HGB was 8.4 g/dl), DIC, portal hypertension, hypersplenism, hematological disorders.

The patient underwent Miles’ procedure (abdominoperineal rectal resection) with sigmoidectomy (fig. 1), en bloc with the uterus and posterior wall of the vagina (fig. 2) and furthermore splenectomy was performed. Intraoperatively marked dilation of rectal venous plexuses extending to the sigmoid colon and the uterus, was found. Due to hematological disorders the patient was transferred in the postoperative period to the Department of Hematology. Histopathology demonstrated: Diffuse cavernous hemangioma of the recto-sigmoid with extraintestinal involvement.

**DISCUSSION**

Due to its low incidence, there are no diagnostic and therapeutic guidelines in patients with suspected DCHR. These are mainly young subjects with recurrent, painless rectal bleeding and anemia (50-75% of patients (1-4, 12,)) as well as less common abdominal pain, constipation and hematemeses (1, 2, 5, 8). Non-specific nature of the symptoms and low incidence of DCHR result in an average delay of diagnosis by approximtely 19 years (1, 6, 7) and numerous “unsuccessful” surgical procedures in this group of patients (1, 4, 6).

Sometimes patients with DCHR undergo diagnostic procedures for hemorrhoids, ulcerative colitis, Crohn’s disease, polyposis, portal hypertension or malignancies (1, 5, 6). First line diagnostic method in suspected DCHR is colonoscopy, allowing for assessment of morphology of the lesion, its location and extent of the infiltration (1, 5, 7). Specimen collection from suspected DCHR lesions is not recommended due to high risk of bleeding (1, 3, 4, 11). MRI is a complementary procedure to colonoscopy. It provides sufficient amount of details that are required to plan the extent of a surgical procedure, enables formation of a multiplane image of a mass as well as imaging without the contrast administration in patients allergic to it (3, 7, 9). CT imaging is characterized by lower sensitivity in diagnosing DCHR, although allows for visualization of calcifications (phleboliths) characteristic for DCHR as well as assessment of extent of the infiltration (2, 3, 7, 9). The other diagnostic procedures (angiography, abdominal ultrasound, plain abdominal X-ray, digital rectal examination) are characterized by low sensitivity and specificity and therefore are not recommended as part of the diagnostic algorithm of DCHR (1, 2, 5, 6, 7, 9).

**Fig. 1.** DCHR – dissected proximal sigmoid colon

**Fig. 2.** DCHR – infiltration of the uterus and posterior vaginal wall
Surgical management of the confirmed DCHR includes an attempt of radical surgical resection of the suspected lesion (1, 4, 6, 7, 9, 10). Due to the fact that DCHR infiltrates the whole width of the rectal wall, minimally invasive techniques, such as endoscopic obliteration, cryotherapy, selective embolization, thermoablation, result only in a transient improvement of the patient’s condition and thus do not reduce the recurrent bleeding rate in the analyzed patient groups (1, 2, 3).

Until 1970 abdominoperoneal rectal resection was the treatment of choice, however due to colostomy unacceptable in this age group and sexual and urological disorders, there has been a tendency to perform this type of procedure only in patients with adverse disease location or infiltration of the perineum, gluteal region or anal canal (1, 3, 10). Attempts of “endorectal pull-through” procedure were reported, although their long term results were unsatisfactory (9). Currently resection of the lesion with sphincter sparing seems to be the treatment of choice – low anterior resection with colorectal anastomosis (1, 5, 6, 9, 10).

Due to the fact that most cases of DCHR originate from linea serrata, this method does not provide complete excision of the lesion, however wide, “sleeve-like” mucosal removal along with the bleeding site provides satisfactory long terms effects and results in marked reduction of complaint rate in patients suffering from DCHR (9, 10, 11).

SUMMARY

Diffuse cavernous hemangioma of the rectum (DCHR) is a rare benign tumor of vascular origin. Diagnostic procedure in patients with DCHR should be based on colonoscopy and MRI. Low anterior resection with colorectal anastomosis is the therapeutic management of choice. Marked delay of the diagnosis in the group of patients with DCHR indicates the requirement for the “selection” of patients with the diagnostic image of DCHR.

REFERENCES