
Diffuse Cavernous Haemangioma of the Rectum: Case Report

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Summary

Diffuse cavernous haemangioma of the rectum, can be misdiagnosed as other common anorectal conditions due to the overlapping clinical presentation and can be caused of massive lower gastrointestinal bleeding leading to significant morbidity. The classic colonoscopic and CT scan findings should be sought and early referral to centres with these investigative modalities should be considered. Treatment includes a complete resection by pull through transection and coloanal anastomosis.

Introduction

Diffuse cavernous haemangiomas of the rectum (DCHR) are rare. Usually confused with anorectal conditions that present with bright red bleeding, its correct treatment is commonly delayed(1). Up to 80% of patients with the DCHR have undergone a minimum of one inappropriate surgical procedure due to misdiagnosis (2). Proper understanding of the condition and careful use of imaging modalities is essential in its early treatment (3). With the paucity of resources in the developing world, the diagnostic challenge is even more daunting. We present a case of DCHR in a 24 year old managed at our hospital. To our knowledge, this is the first reported case of DCHR in the East African literature.

Case Report

A 24 year old female presented with a five month history of recurrent fresh and painless rectal bleeding, occasional passage of clots and had a sensation of incomplete rectal emptying. She denied melaena, haematemesis or mucosal bleeding. There was no history of abdominal pain, altered bowel habits (no constipation or diarrhoea) or weight loss. Her past surgical history was significant for several transfusions for severe anemia, hemorrhoidectomy for rectal bleeding, left hemicolectomy with primary anastomosis one year earlier for massive lower gastrointestinal bleeding requiring seven units of blood transfusion. She had no family history of malignancy. She was a college student with no history of smoking and only took alcohol occasionally.

On first encounter at our institution, she was very pale, not dyspnoeic or jaundiced, the patient was well hydrated, had no generalized lymphadenopathy or lower limb oedema. The blood pressure of 100/70 mmHg, a pulse rate of 84 beats per minute and temperature of 36.8°C. She was fully oriented and had a normal respiratory and cardiovascular examination. Abdominal examination revealed a non-tender abdomen, a well-healed laparotomy scar and no palpable masses. Digital rectal examination revealed a normal anal tone, smooth anorectal mucosa, no masses, and soft stools which were non-bloody. Anoscopy was essentially non-revealing. Her haemoglobin level was 3.6mg/dl and she received four units of packed cells at admission. The coagulation screen was normal. She was prepared for colonoscopy and the findings are shown on Fig. 1. Bleeding continued after colonoscopy necessitating transfusion of one more unit of packed red blood cells. A CT scan abdomen done revealed the typical thickening of the rectal wall with multiple serpentine vessels and phleboliths (Fig 2a-c).She was counselled for low anterior resection and coloanal anastomosis. After adequate colonic preparation, her pre-operative haemoglobin was 10.8g/dl and three more units of packed cells were available for surgery. Through a midline abdominal incision, the pelvis was explored, rectum mobilised and an arteriovenous malformation (AVM) extending from the rectosigmoid junction to the anorectal junction was found. The AVM was carefully excised and hand-sewn coloanal anastomosis done with a diverting loop transverse co-

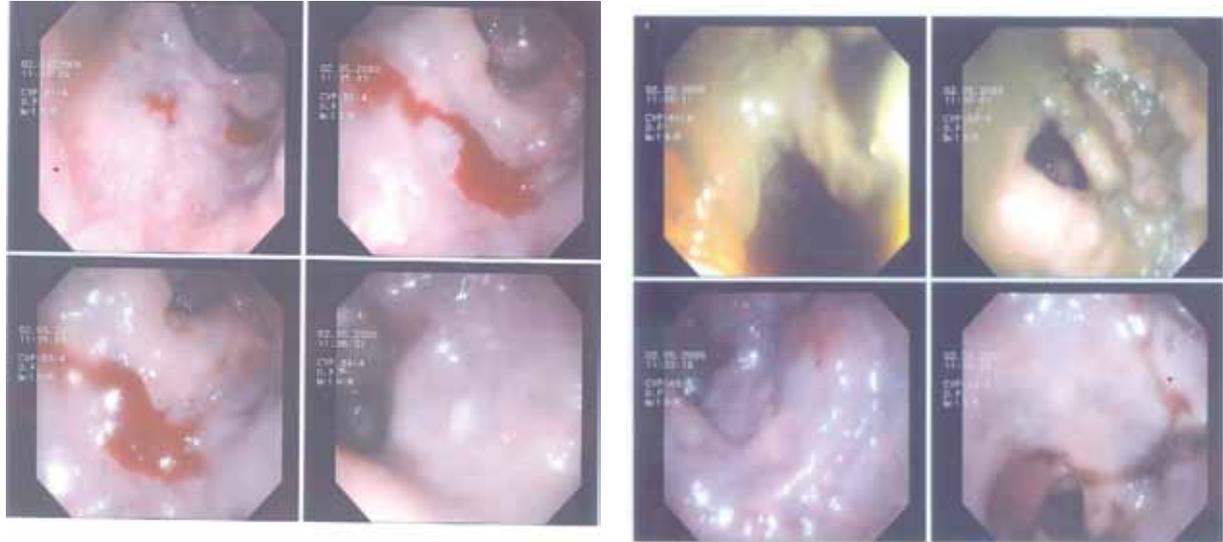


Fig 1: Colonoscopic images of the patient showing multiple bluish - purple submucosal masses representing the dilated tortuous blood vessels.

lostomy fashioned. Total operative time was four hours and total blood loss was 250mls. She had an uneventful post-operative period and was discharged on the 9th postoperative day.

On follow up in the surgical outpatient clinic, a gastrograffin enema revealed stenosis at the anastomosis and rectovaginal fistula 1.5cm from the anal verge. The latter has subsequently closed spontaneously and she is due for colostomy closure. The stenosis responded to serial anal dilatations.

The surgical specimen (Fig 3) was sent for histology and confirmed the diagnosis of Diffuse Cavernous Haemangioma of the Rectum.

Discussion

Diffuse Cavernous Haemangioma of the Rectum (DCHR) is a rare benign lesion described in all age groups and may present with severe bleeding per rectum. It was first described by Philips in 1839 since when about 100 cases of DCHR have been reported (4). It may mimic internal haemorrhoids, ulcerative colitis, carcinoma, and adenomatous polyp in patients who present

with a history of intermittent painless bright red rectal bleeding. Hence, a misdiagnosis is not uncommon (1). The delay time between initial symptoms and final diagnosis ranges from 8 to 50 years(5). The mean delay time between initial symptoms and final diagnosis was 17.63 years (range = 0–48 years) in one series of 17 patients (6) where seven patients had been misdiagnosed as having haemorrhoids with one undergoing at least seven haemorrhoidectomies(6). In the developing world, the challenges are amplified due to the lack of adequate resources to properly investigate this condition. Our case illustrates these challenges as the patient had undergone both a haemorrhoidectomy and left hemicolectomy, with the diagnosis being made much later after referral to a center with investigative resources.

The submucosal vascular plexuses that form the colonic hemangiomas, result from embryonic sequestration of mesodermal tissue. Histologically distinct from telangiectasias and angiodysplasias, hemangiomas are classified as capillary, cavernous, or mixed(1). Cavernous hemangiomas, which comprise 70% of colonic haemangiomas, may occur as part of multisystemic or diffuse

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Fig 2: CT scan images of the patient shows thickening of the rectal wall with narrowing of the lumen filled with water contrast (a). Tortuous blood vessels representing serpentine enlarged blood vessels in the distal rectum(b), and characteristic calcifications in the rectal wall (c).



Fig 3: Surgical pathology specimen from the patient confirming the diagnosis of diffuse cavernous haemangioma involving the rectum with marked, focal submucosal haemorrhage and multiple organizing thrombi involving the submucosa, serosa, and mesenteric fat.

gastrointestinal angiomatosis, with the rectosigmoid as the most common site of occurrence (7).

Even though proctosigmoidoscopy may confirm the diagnosis in some cases, a careful biopsy with its potential hazards of causing profuse haemorrhage may remain the only option in most others. Characteristic endoscopic findings are of soft, dilated, easily collapsible submucosal masses, ranging in color from deep wine to plum (8). Computed tomography (CT) scans can give an accurate diagnosis, showing a thickened rectosigmoid colon and typical pelvic phleboliths, and possibly, the extrarectal lesions (9). In addition, it can provide a pertinent preoperative information including longitudinal extent of bowel involvement, degree of bowel wall thickening, involvement of adjacent pelvic viscera, and multiplicity and vascularity of the lesion(3). Abdominal radiography which might at times be the only available modality in resource poor settings may reveal the characteristic multiple pelvic phleboliths but the positive rate of abdominal radiography can be as low as 26–50% in adult patients (5, 6).

Due to its origin from the dentate line and the involvement of the whole layer of the rectal wall and the rectal mesentery, the treatment of choice for DCHR is complete resection by pull-through transection and coloanal anastomosis(6). With the development of surgical skills and the application of the double-staple technique, low anterior resection (LAR) was advocated for the treatment of DCHR (8).

In conclusion, DCHR is a rare condition that may mimic other causes of lower gastrointestinal bleeding. It must be considered in the differential diagnosis of recurrent rectal bleeding and adequate and timely investigations done in order to institute the correct treatment.

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