

IMAGING FINDINGS IN DIFFUSE RECTAL CAVERNOUS HEMANGIOMATOSIS: A RARE CAUSE FOR PER RECTAL BLEEDING

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Keywords : *rectal cavernous hemangiomas, rectal bleeding*

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Background

Cavernous hemangiomas of the colon are a rare benign entity which is important to be recognized by radiologists during imaging, in order for surgeons to be able to avoid biopsy which could in turn lead to catastrophic bleeding. Patients usually present with painless per rectal bleeding, in their youth¹. It has an equal sex distribution¹. Majority of patients undergo a wide variety of investigations and are commonly misdiagnosed. The average time elapsed from presentation to diagnosis is reported in recent literature as 17.63 years².

Case Presentation

A 59 year old female, with multiple co morbidities, presented to us with a history of intermittent painless per rectal bleeding since 12 years of age. She passes a few drops of blood at the end of defecation. She did not notice any palpable lumps. There were no episodes of torrential bleeding at any point. Since childhood, her bowel movements were regular, once in two days. There was no alteration of bowel habits. On digital rectal examination, there were no palpable mass lesions. She did not have any cutaneous manifestations of congenital vascular anomalies. She had not undergone any previous imaging in evaluation for per rectal bleeding during her childhood. There was no family history of congenital vascular malformations or bowel malignancies.

The possibility of a vascular malformation was first questioned after she underwent a colonoscopy at 30 years of age. There were sub mucosal dilated bluish lesions involving the rectum, with no evidence of synchronous mass lesions. She chose to be managed conservatively with serial colonoscopies. Her haemoglobin levels were serially monitored, which was maintained at no less than 12g/dl throughout. The last colonoscopy was done in early 2020, after a surveillance interval of 5 years, which showed progression of the possible vascular malformation, now involving the anus and rectum extending up to the recto sigmoid junction. However, her clinical symptoms were none progressive.



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She was referred to our unit for a Contrast Enhanced Computed Tomography (CECT) of the abdomen and pelvis region. The study was done on a Toshiba Aquilion 16 slice machine with intravenous contrast (Omnipaque) 1ml/kg dose, with rectal and oral water. There was eccentric bowel wall thickening involving the anal canal and rectum with maximum thickness in the mid rectum. The diffuse wall thickening extended to involve the rectosigmoid junction and distal part of the sigmoid colon with an abrupt transition to normal bowel loops proximally. There were multiple peri rectal phleboliths throughout the affected length. There was normal mucosal enhancement with no abnormal draining or feeding vessels identified within the walls of the affected segment. There was extensive peri rectal fat stranding with widening of the pre sacral space.

This was followed by a Magnetic Resonance Image (MRI) of the pelvis on a 3 Tesla machine. T1W, T2W axial and coronal sequences, T2W sagittal, T2W FS axial with dynamic contrast enhanced images were acquired in a 3 Tesla (3T) machine. MRI showed characteristic T2 high signal intensity within the rectal wall with extensive wall thickening. Signal voids in T2W images were seen within the rectal wall, compatible with phleboliths. There were no dilated regional veins or arteries and no evidence of extrarectal extension and invasion of adjacent structures.

Imaging findings were suggestive of diffuse cavernous hemangiomas of the rectum (DCHR). She opted for conservative management with a close follow up plan, as her symptoms were non progressive.

Discussion

Colonic hemangiomas are rare benign vascular lesions mainly arising from the sub mucosal vascular plexus³. Histologically they can be categorized in to capillary, cavernous, arteriovenous and mixed types⁴. 80% of colonic hemangiomas are cavernous hemangiomas containing large thin walled vascular structures. Cavernous hemangiomas commonly involve the recto sigmoid junction and can be characterized as discrete or diffuse involvement. Extensive diffuse involvement of the rectum is known as diffuse cavernous hemangiomas of the rectum (DCHR) and is considered a rare entity³. DCHR originates commonly from the dentate line and the distribution is mainly limited to the rectosigmoid region. Rectal involvement with extension of the disease to the anal canal is unusual⁵. There are no reports in literature to suggest an association between DCHR and colorectal malignancies. However, there are isolated case reports of coexisting tubular adenomas, infiltrative adenocarcinomas^{6,7}.

Histologically, DCHR contain large thick walled vascular channels lined by single or multiple layers of endothelial cells⁸. Stasis of blood flow, leading to thrombus formation and subsequent chronic inflammation leads to phlebolith formation. Full thickness mural involvement is usually common and there are cases reporting invasion of adjacent structures such as gluteus maximus muscle and lower limb musculature².

DCHR mainly presents in the youth with equal sex predilection¹. Majority of patients will be asymptomatic. Painless per rectal bleeding is the commonest presenting symptom⁹. In addition, patients can also present with iron deficiency anemia, intestinal obstruction and bowel perforation³. Colonic haemangiomas are associated with conditions such as Klippel Trenaunay and Osler Weber Rendu syndrome and can have associated cutaneous manifestations. However, in DCHR cutaneous manifestations are considered uncommon.

In colonoscopic assessment, these lesions appear as submucosal hypervascular lesions, bluish purple in color, compressible and protruding in to the lumen. This appearance can simulate hemorrhoids, varices or polyps. Biopsy, if mistakenly performed can be catastrophic for the patient. Therefore, radiological evaluation is optimal for DCHR due to the availability of non invasive, imaging modalities which shows characteristic imaging findings hence negates unnecessary biopsies.

CECT is an effective diagnostic method of the condition. Characteristic appearances in CT were observed in our patient, which included diffuse, circumferential, enhancing, transmural wall thickening with multiple phleboliths within the bowel wall. Associated peri rectal fat stranding and widening of the pre sacral space was also seen in our patient. The primary advantage of CT over endoscopic evaluation is that, transmural wall thickening can be accurately assessed. This in turn is crucial to exclude mimicking lesions in endoscopy³. However, radiation exposure and the use of iodinated contrast agents can be drawbacks.

MRI is considered as the gold standard imaging method in the evaluation of DCHR^{10, 11}. It's soft tissue resolution and contrast resolution are superior when compared to CT. It also helps decipher the extent of involvement better than in CT¹¹. Characteristically, rectosigmoid wall thickening with high signal intensity, grape like lobulated lesions are seen on T2-weighted images¹⁰. Areas of signal void within the walls are seen indicative of phleboliths. Usually dilated regional arteries or veins are not seen¹¹. The extent of bowel involvement and extra rectal locations are well shown on T2-weighted MR images¹¹. These constellation of imaging findings are not seen in other rectal pathologies and are considered more specific than CT findings of DCHR¹¹. Thus MRI is an important non invasive and accurate diagnostic tool.

Surgical resection is considered as the definitive treatment for symptomatic patients. Sclerotherapy, embolization, electrocautery are considered other management options³.

In conclusion, the presence of typical imaging findings in CT and MRI, leads us to a confident and accurate diagnosis of DCHR. It should be considered as a rare differential diagnosis of painless per rectal bleeding at any age group. The early diagnosis of the condition can enable surgeons to avoid biopsies and proceed with definitive treatment.

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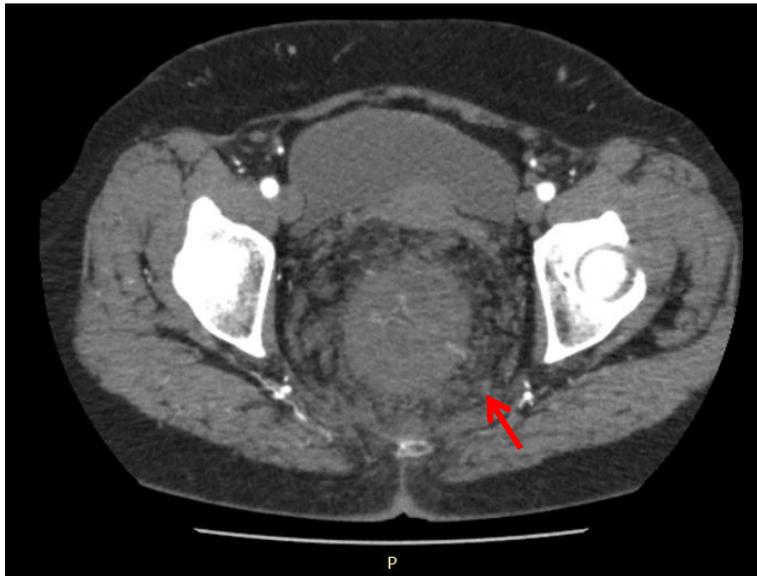


Figure 1: Axial image in the Non contrast phase showing extensive peri rectal fat stranding with diffuse thickening of the rectum (Arrow)

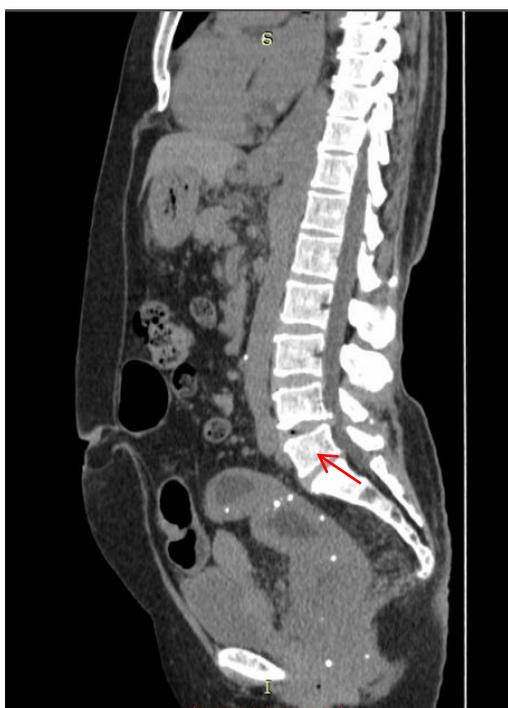


Figure 2: Sagittal image showing peri rectal phleboliths(Arrow) within the rectal wall and widened pre sacral space.

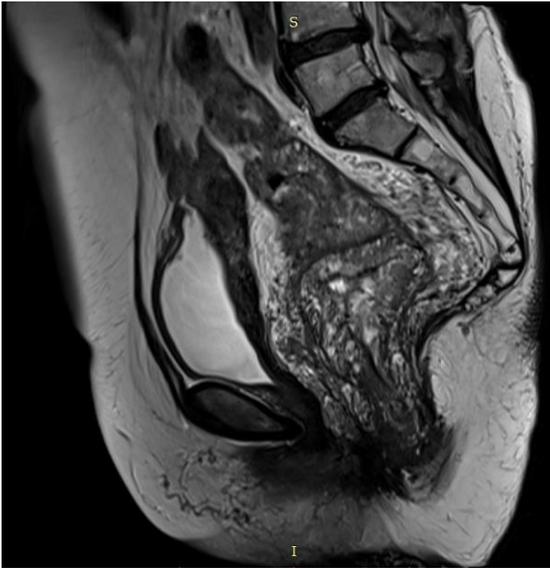


Figure 3: T2 weighted sagittal image showing T2 signal voids characteristic of phleboliths

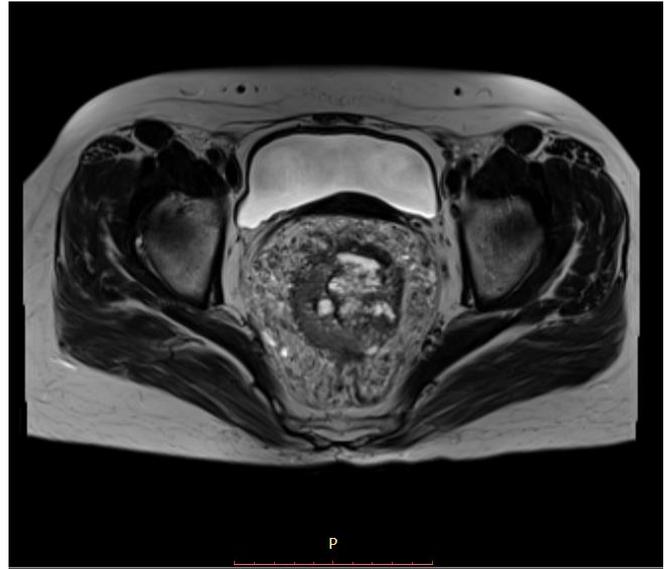


Figure 4: T2 weighted axial image demonstrates extensive rectal wall thickening

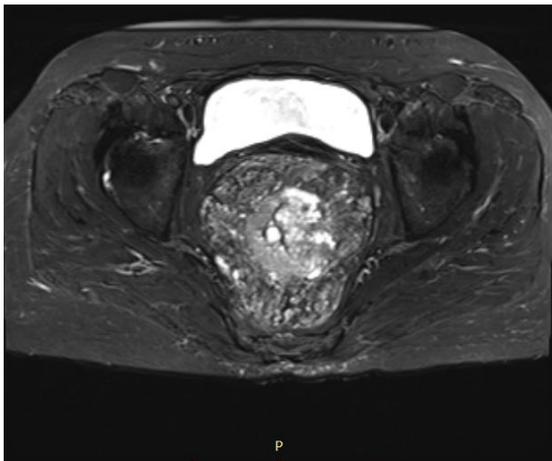


Figure 5: T2 STIR axial image shows T2 high signal intensity with rectal wall thickening

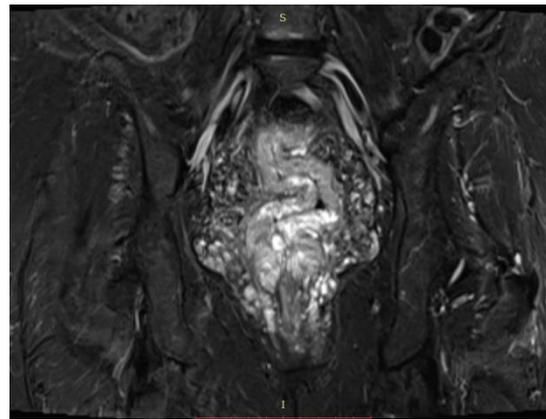


Figure 6: T2 STIR coronal image depicts the thickened T2 high signal rectal wall