



# A pictorial review of rectosigmoid cavernous vascular malformations

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# Learning Aims



UNDERSTAND THE  
PATHOPHYSIOLOGY AND ITS  
ASSOCIATED NOMENCULTURE



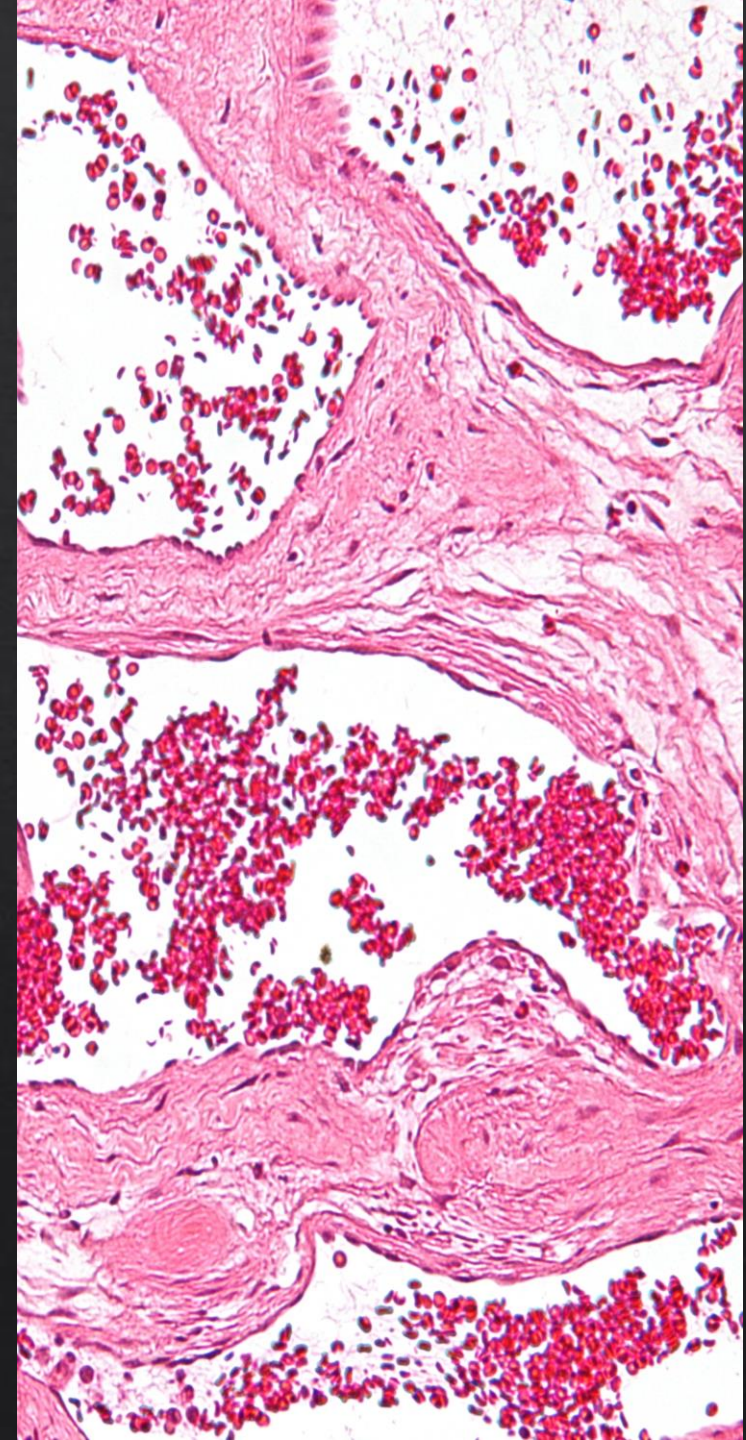
INCREASE FAMILIARITY WITH  
THE X- RAY, CT AND MRI  
FINDINGS.



APPRECIATE THE DIFFERENT  
MANAGEMENT OPTIONS  
AVAILABLE

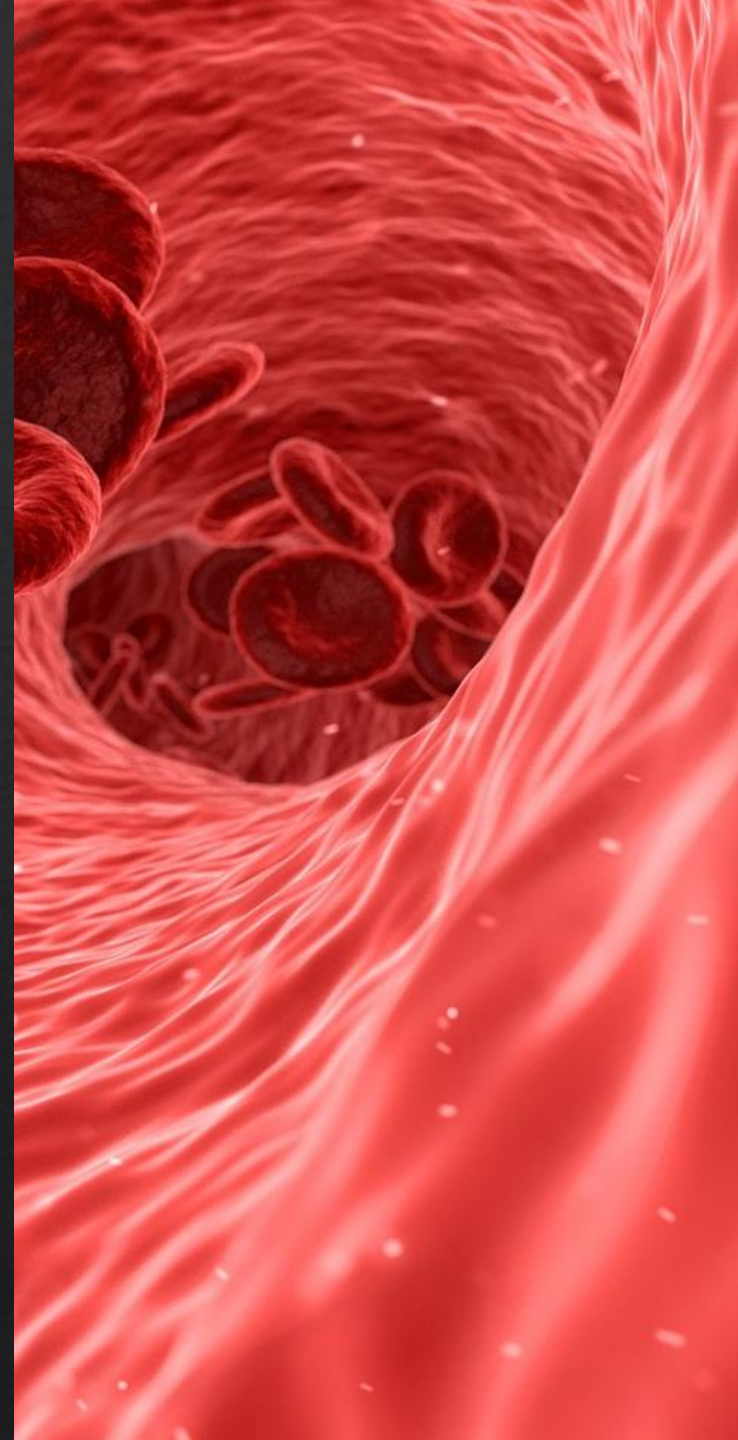
# Pathophysiology

- ◇ While these lesions are commonly referred to as haemangiomas they are better characterized as venous malformations (1)
- ◇ 80% of rectosigmoid malformations are the cavernous subtype. They can be either of local or diffuse variety (2).
- ◇ On histology they show large spaces lined by endothelial cells. These have formed due to embryological lack of smooth muscle, causing enlarging spaces from hydrostatic pressures . This contrasts with true haemangiomas which enlarge by cell proliferation (3).
- ◇ These lesions are unusual in their ability to invade local structures.



# Symptoms

- ◆ The first symptoms often occur in childhood.
- ◆ Most patient present with per rectal bleeding and can have associated chronic iron deficiency anemia (1).
- ◆ Other presenting symptoms include obstruction secondary to circumferential masses, polypoid abnormalities acting as lead points, pelvic pain, tenesmus and constipation.



# Diagnostics

- ◆ Unfortunately, delayed diagnosis is common.
- ◆ In one case series of 17 patients there was a mean delay between initial symptoms and diagnosis of 17.5 years.
- ◆ Patients are mistaken for having haemorrhoids, colitis, portal hypertension and rectal polyps. This has led to unnecessary interventions including haemorrhoidectomies, polypectomies, transfixation and sclerotherapy for rectal varices and being given mesalazine for colitis (1).



# CT Imaging

- ◆ On CT the pathognomonic findings are inhomogeneous enhancing transmural bowel wall thickening with phleboliths.
- ◆ CT will be more sensitive for identification of the phleboliths than plain film. It can prove they exist within the abnormal lesion as opposed to incidental venous phleboliths within the pelvis.
- ◆ CT allows for proof of the transmural nature of the cavernous malformations and helps to exclude differentials such as mucosal inflammatory processes.
- ◆ CT virtual colonoscopy has also been used with suggested benefits of noninvasive double contrast barium enema views and subsequent assessment of the mucosal detail (1).

# MRI

- ◇ MRI has added benefits in rectal cavernous malformations. It can show the increased T2 signal within the bowel wall, thought due to slow flow.
- ◇ There can also be adjacent high T2 signal in the peri-rectal fat.
- ◇ Serpiginous structures also correlate with the small vessels in the malformation.
- ◇ Hemorrhoids can be distinguished due to the lack of perirectal fat high T2 and the higher location.
- ◇ MRI can aid in surgical planning, in advising the length of normal remnant rectum for reconstruction and sphincter involvement, transferable skills learnt from rectal malignancy assessment (1).

# Management

- ◆ Surgical resection is considered definitive treatment, ideally with a sphincter saving approach (1).
- ◆ This may not be appropriate in all patients and many different treatment options have been employed.
- ◆ This can include alternative interventions such as endoscopic resection, sclerotherapy and cryotherapy. (2).
- ◆ Medical therapies have also been used with reported success, from tranexamic acid (3), to propranolol, celecoxib and thalidomide due to their action of vascular endothelial growth factor (4).



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3. Cotzias E, Rehman SF, Arsalani Zadeh R, Smith D. Conservative management of diffuse cavernous haemangioma of the sigmoid and rectum. *Ann R Coll Surg Engl*. 2020 Jan;102(1):e1-e3. doi: 10.1308/rcsann.2019.0088. Epub 2019

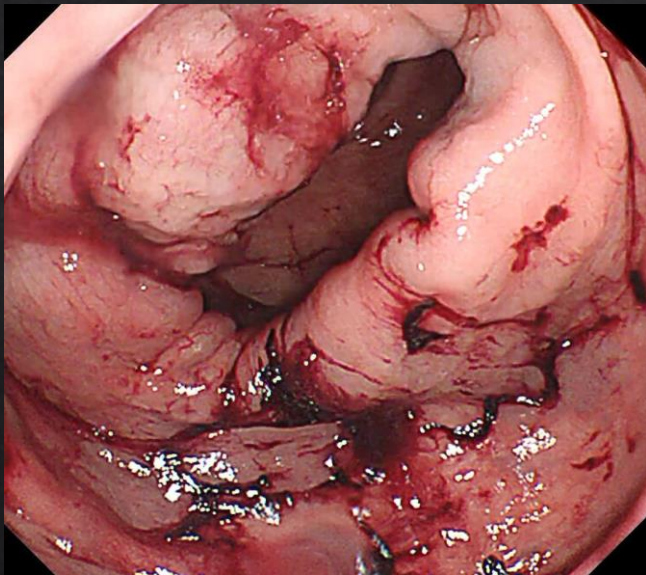
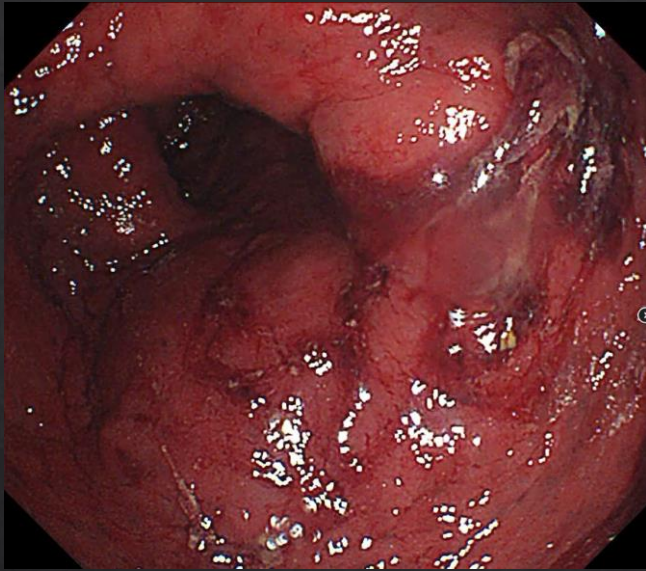
4. Abematsu, Takanari & Okamoto, Yasuhiro & Nakagawa, Shunsuke & Kurauchi, Koichiro & Kodama, Yuichi & Nishikawa, Takuro & Tanabe, Takayuki & Shinkoda, Yuichi & Mukai, Motoi & Kaji, Tatsuru & Kawano, Yoshifumi. (2015). Propranolol and celecoxib. *Journal of Pediatric Surgery Case Reports*. 3. 331-333. 10.1016/j.epsc.2015.06.009.



# Case review

# Case 1

- ◇ 66-Year-old female patient
- ◇ PC: PR bleeding and diarrhoea. Intermittent PR bleeding since childhood.
- ◇ PMH: AF, hypertension, COPD, Asthma, T2DM, Schizophrenia, Arthritis
- ◇ O/E: Distended abdomen, tender all over. Dark red blood on PR exam.
- ◇ Bloods: Hb 90, WCC 5.6, Plt 263, EGFR >90
- ◇ Review of notes show history and investigations dating back 8 years prior, where the diagnosis had been made.



Endoscopy at the time showed : Rectum and distal sigmoid with abnormal nodular congested mucosa, friable on touch of the scope and bleeding on biopsies. Submucosal oedema and hematomas.

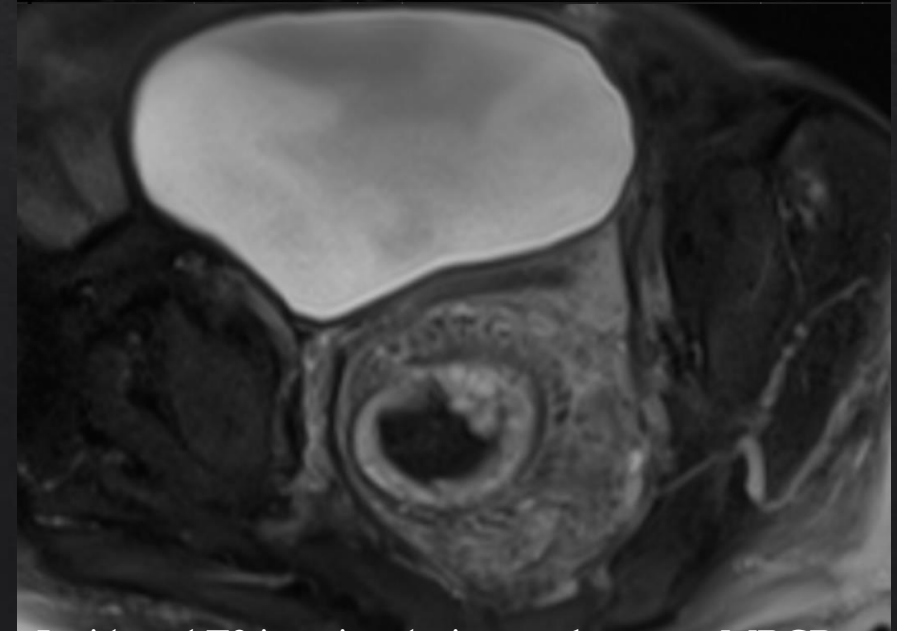
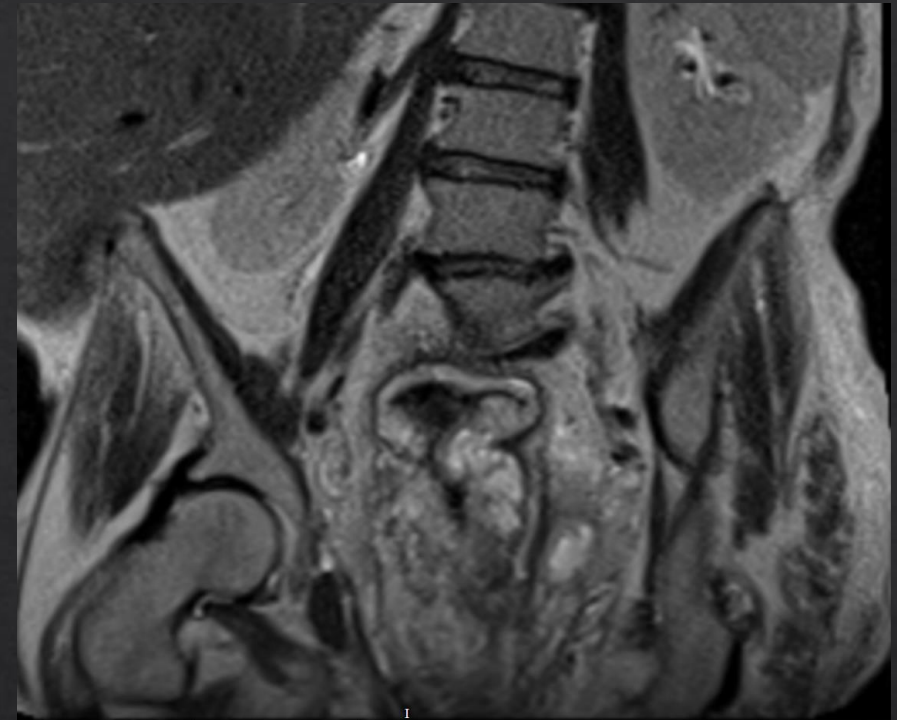


CT performed 3 months prior:  
There is diffuse circumferential transmurial wall thickening (white triangles) involving the rectum and sigmoid colon with multiple calcific densities within it representing phleboliths (white arrows).

# Incidental imaging during the patients care.



Pelvic Xray: There are diffuse calcific densities within the pelvis in keeping with phleboliths.



Incidental T2 imaging during a subsequent MRCP showing high signal within the bowel wall.

# Case 2

- ◇ 39 Female
- ◇ PC: Pelvic pain. This had been thought to be secondary to a Bartholin's cyst. She has had a previous excision of a right labial Bartholin's cyst.
- ◇ PMH : Previous examination and excision of a right sided vulvovaginal haemangiomatic mass 15 years prior under anesthesia. Previous partial clitorrectomy due to painful thrombosis in the region. Recurrent right Bartholin's cysts requiring surgical excision.
- ◇ Observations normal
- ◇ O/E perineal mass like lesion. True cut biopsy performed.
- ◇ Histology: Sections of fibrous tissue containing thin-walled dilated vascular channels. The appearances are those of a haemangioma.

# MRI rectum for surgical planning



MRI Pelvis and rectum: There was space demanding high T2 signal within the mesorectal fascia. This can be seen to be continuous with the rectal wall (white arrows) in the upper rectum and separate from the upper vagina (white triangles). There is extension of the lesion through the inter-sphincteric plane and into the perineum. The signal abnormality extends anterior within the perineum (blue arrow) and involves the right labia majora (purple arrow).



Repeat MRI 10 years later showed long term stability in the size and degree of invasion.



CT performed with the view to arterial embolization. Typical calcification within the lesion. No target identified.

# Case 3

- ◇ Legacy case
- ◇ 62-year-old female
- ◇ Limited history available.
- ◇ Investigated for acanthosis nigricans
- ◇ CT Enema performed.





Left: MIP coronal CT images show the typical phleboliths within the pelvis, related to the thickened colonic wall.  
Above: Long segment rectosigmoid bowel wall thickening with mural phleboliths.  
Right: 3D reformats showing the pelvic phleboliths.

# Take home points

- ◆ These lesions are not a true haemangioma but vascular malformations
- ◆ The characteristic appearances on CT of transmural bowel wall thickening and phleboliths.
- ◆ MRI's added use in surgical planning for sphincter assessment and distal anastomosis planning

Thank you.