

# Venous and lymphatic vascular hamartoma of the recto sigmoid: a rare case of paediatric per-rectal bleeding

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### Introduction

Hamartoma is a benign tumour like malformation that manifest during normal tissue development. The size of the hamatoma increases in relation to the growth of the body volume and it is unlikely to involute [1]. Vascular hamartoma includes haemangioma, lymphangioma and arterio - venous malformation.

Vascular lesions of the Gastro intestinal tract, although an infrequent occurrence [2], should be investigated. The clinical triad of painless rectal bleeding, phleboliths on imaging and cutaneous lesions points towards a diagnosis of a vascular lesion [3]. Based on histopathological analysis which showed a venous and lymphatic hamartoma, the presented case is a rareness in the literature [4].

### Case report

An 8 year old girl was admitted with haematochezia and tenesmus not associated with loose stools or mucus diarrhoea. She had previously been investigated at one year but no diagnosis was made. The symptoms too had resolved spontaneously from three years of age until this presentation.

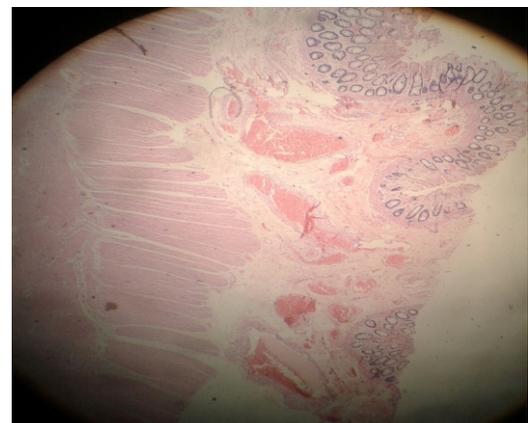
She was severely anaemic with a haemoglobin of 5.7 g/dl with fresh bleeding per rectum with a normal coagulation profile and inflammatory markers. An urgent Lower GI endoscopy revealed a distinctly inflamed rectal wall extending from the anal verge until the recto sigmoid junction. A trial of oral steroids and steroid enema (Enterfoam®) was commenced due to the strong clinical suspicion of an Inflammatory Bowel Disease (IBD). Subsequently, Magnetic Resonance Imaging (MRI) revealed a vascular malformation in the rectal wall extending from S4 level to the anal canal and extending into the ischio-rectal fossa. An urgent abdominal Contrast Enhanced Computed Tomography (CECT) was done next which also demonstrated several coarse calcifications (phleboliths). Following the

radiological 'suggestion of vascular malformation a Digital Subtraction Angiography (DSA) was performed. However no definitive arterio-venous malformation was seen. Despite a negative DSA, an on-going life threatening haemorrhage was taken into account at a Multi-Disciplinary meeting and an urgent laparoscopic pull through surgery was planned.

The surgery showed dilated venous plexus around the rectum with dilated veins in the sub-serosa of the rectum. Histology revealed a lymphatic and vascular sub-mucosal hamartoma.



**Figure 1.** CT scan reveals significantly thickened recto-sigmoid wall with phleboliths (star)



**Figure 2.** Blood containing sinuses with prominent lymphatics and proliferating vessels with intramural thrombi in the sub mucosa.

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## Discussion

In the evaluation of painless per rectal bleeding in children polyps, Inflammatory Bowel Disease and Meckels diverticulum are more frequently encountered over vascular lesions, yet their existence should not be far off down the list.

The nomenclatures of vascular lesions have been confusing leading to a difficulty in identifying and treating them. The latest classification is by the International society for the study of vascular anomalies (ISSVA); the classification system divides vascular anomalies into vascular neoplasms and vascular malformations.

A haemangioma has a high endothelial cell turnover and are further sub-classified into infantile haemangioma, rapidly involuting congenital haemangioma (RICH), non involuting congenital haemangioma (NICH), Kaposiform haemangiioendothelioma, and tufted angioma. One third of haemangiomas are present at birth. The rapid growth in the first two years is followed by involution in 90% of cases. Steroid administration accelerates the involution phase although propranolol is the gold standard of treatment. Vascular malformations are considered to be structural abnormalities of the capillary, venous, lymphatic and arterial systems which increase in size with the growth of the child. However they do not have increased endothelial cell turnover, are always present at birth and involution is not a character.

A vascular hamartoma incorporates both these components. It is classified as an abnormally arranged excessive tissue in the tissue of its origin which grows in relation to the body size with no evidence of spontaneous involution. Thus it is defined as a benign vascular neoplasm of unknown aetiology with no strong genetic predisposition.

## Diagnostic difficulty

Over 80% of patients with vascular lesions undergo at least one inappropriate surgical intervention prior to diagnosis. Painless hematochezia and iron deficiency anaemia are the most frequently encountered. Tenesmus, pelvic pain, intermittent abdominal pain, intestinal obstruction, intussusception and perforation are also seen.

An upper GI endoscopy needs to be done to exclude upper GI bleeding and a single colonoscopy is crucial. The pathognomonic finding is the blue to red intra luminal projections with or without active bleeding points and ulceration.

An abdominal radiograph may demonstrate the presence of calcified phleboliths. Barium studies are useful in obstructive polypoid lesions but vascular lesions may be compressed during air insufflation.

In our patient the Magnetic Resonance Imaging (MRI) scan

revealed a vascular malformation in the rectal wall and computed Tomography (CT) further supported the diagnosis by the presence of trans-mural enhancing bowel wall thickening with the presence of characteristic phleboliths. Magnetic Resonance Imaging (MRI) findings is more useful for the diagnosis of rectal vascular lesions. However due to diagnostic dilemma and persistent bleeding, we had to perform Contrast Enhanced Computed Tomography (CECT) for this patient. Endo rectal ultrasound with Doppler flow studies is another imaging modality which may have been useful in diagnosing if paediatric probes are available.

Following strong clinical and radiological evidence we proceeded to mesenteric angiography for diagnosis and treatment. However the negative angiography result may be either due to the presence of thrombosis or slow flow venous lesions.

Management of paediatric rectal bleeding warrants for urgent resuscitation. In a pure haemangioma, steroids have been proven to be successful but many GI haemangioma consist of partial vascular malformations. This explains the cessation of rectal bleeding following rectal steroid enema initially but which recurred due to the vascular hamartoma of venous and lymphatic origin.

Endoscopic resections are used for more proximal and smaller lesions. In this case a laparoscopic sphincter saving pull-through surgery was chosen because of the severity of hematochezia and failed DSA. Therefore in most cases the final diagnosis is established through surgery and histopathology.

The patient remains asymptomatic at the time of writing.

## Conclusion

Vascular lesions must be considered when evaluating paediatric GI bleeding. Diagnosis can be made by thorough clinical history taking, imaging by experience radiologist, and supportive endoscopy. Precise use of terminology should be emphasized with reference to the latest guidelines and classifications. Surgery continues to play a key role in the definitive care and minimally invasive techniques are used to manage recurrences.

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**Key Points:**

- Vascular hamatoma in rectum is a rare cause for paediatric per rectal bleeding however it can lead to life threatening bleeding if not treated.
- Diagnosis of hamatoma is difficult due to lack of experience and resource materials.
- Focus radiological assessment by an experience radiologist is pivotal and multidisciplinary surgical approach is essential.