

Unusual cause of 55 years of rectal bleeding: hemolymphangioma (a case report)

Sagar Pandey, MD^a, Miao Fan, MD, PhD^{a,*}, Junfeng Zhu, MD, PhD^b, Xiaofang Lu, MD^b, Dandan Chang, MD, PhD^a, Xiuhong Li, MD, PhD^{c,*}

Abstract

Rationale: Hemolymphangioma is a rare developmental error of combined blood and lymphatic vasculature. To the best of our knowledge, there is only one case of rectal hemolymphangioma reported in Pubmed. Our case probably is the first reported rectal hemolymphangioma with computed tomography (CT) evaluation.

Patient concerns and diagnosis: A 57-year-old male was presented to our hospital with 55 years of long history of episodic rectal bleeding. Past medical history showed numerous hospital visits for similar illness. Multiple diagnoses were made and different treatment modalities were applied for his benefit, but none of them relieved the symptoms permanently. He was then referred to our hospital. On admission, he was presented with intermediate rectal bleeding of fresh blood. CT examination showed isodense homogenous rectal wall thickening with heterogeneous enhancement on contrast examination. Multiple calcifications were seen in and around the lesion.

Interventions and outcomes: He underwent open abdominal surgery with total surgical excision of the lesion. Post-surgical histopathological examination of excised specimen showed submucosal multiple thin-walled vessel of varying size, some consistent with blood vessel and other with lymph vessel, thus diagnosis of hemolymphangioma was made. Follow-up for 6 months showed no recurrence.

Lessions: Hemolymphangioma is a benign developmental lesion. Radiological findings can be challenging and range from benign cystic lesion to aggressive lesion mimicking malignancy. Therefore, combined clinical history, radiological findings, and continuous follow-up can help make proper diagnosis and provide prompt and accurate treatment.

Abbreviation: CT = computed tomography.

Keywords: computed tomography, hemolymphangioma, lymphatic malformation, venous malformation

1. Introduction

Hemolymphangioma is a rare developmental defect of combined blood and lymphatic vasculature.^[1] It is believed to occur due to somatic mutation in developing vascular tissue. The coexistence of these 2 abnormal vasculatures in hemolymphangioma is not well understood, but it strengthens the hypothesis that both blood and lymph-endothelial cell originate from common stem cell. As the lesion consists of 2 different vascular tissues with interposed stroma

at varying proportion, radiological findings can vary accordingly.^[2] Only few cases of hemolymphangioma have been reported. To the best of our knowledge, there is only 1 case of rectal hemolymphangioma reported in PubMed.^[3] Probably ours is the second case of rectal hemolymphangioma, but the long history of clinical presentation and involvement of multiple organs is special than the earlier reported case. Also, our case probably is the first reported rectal hemolymphangioma with computed tomography (CT) evaluation. This essay is a case report of a rectal hemolymphangioma in a 57-year-old male. The clinical features, radiological, and pathological findings of the case are also discussed.

2. Case report

A 57-year-old male was admitted to our hospital with 5 months' history of massive rectal bleeding (rectorrhagia). He had past history of passage of fresh blood mixed stool since last 55 years. He first complained of rectal bleeding at the age of 2. He went hospital numerous times for this symptom; multiple interventions were done but disease was not cured completely. During one of his hospital visit in the past (patient forgot the date), he was diagnosed and treated as hemorrhoids. The symptom was relieved for a while and then recurred again. Thirty years ago, he was diagnosed as rectal hemangioma and managed with cryotherapy in our hospital. That could also just relieve the symptom for few years and it recurred again. Then, he consulted many other hospitals, but could only get short-term symptomatic relief without proper treatment of the cause. Since 5 months, the severity of rectal bleeding increased. He then went to local hospital where intervention was done to control bleeding and

Editor: Farid Azmoudeh-Ardalan.

The authors report no conflicts of interest.

^a Department of Radiology, ^b Department of Pathology, ^c Department of Maternal and Child Health, School of Public Health, Sun Yat-sen University, Guangdong, China.

* Correspondence: Miao Fan, Department of Radiology, the First Affiliated Hospital of Sun Yat-Sen University, No.58, Zhongshan 2nd Rd, Guangzhou, Guangdong 510080, China (e-mail: cmu-sums@163.com); Xiuhong Li, Department of Maternal and Child Health, School of Public Health, Sun Yat-sen University, No.74, Zhongshan 2nd Rd, Guangzhou, Guangdong 510080, China (e-mail: lixh@mail.sysu.edu.cn).

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Medicine (2017) 96:10(e6264)

Received: 25 August 2016 / Received in final form: 7 February 2017 / Accepted: 8 February 2017

<http://dx.doi.org/10.1097/MD.0000000000006264>

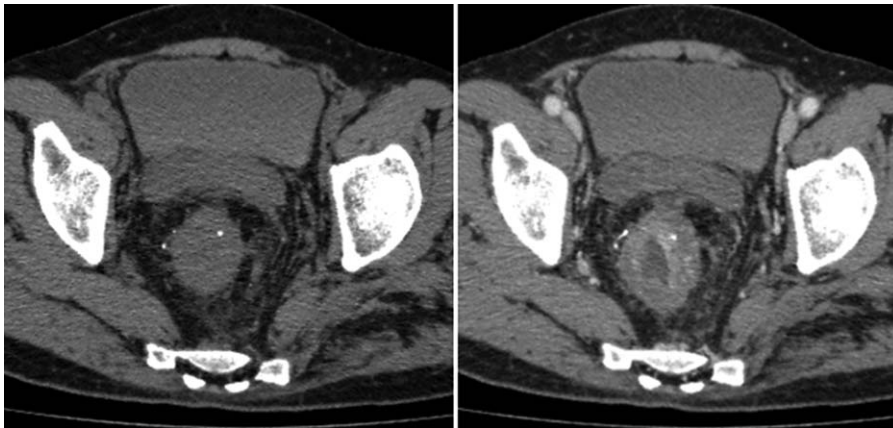


Figure 1. Axial view of pelvis. Axial view at pelvis showed homogeneously thickened rectal wall on none enhanced CT (left), which showed heterogeneous enhancement on venous phase contrast enhanced (right). Multiple small calcifications are seen in and around the bowel wall.

symptomatic treatment was done with intravenous fluid and blood transfusion. He finally referred to our hospital for further evaluation.

Intermediate rectal bleeding of fresh blood was presented on admission. Patient complained of dizziness on standing, shortness of breath, and palpitation on walking a short distance. There was no history of similar illness in family. On general examination, patient appeared pale, but his heart rate and blood pressure were within normal limit during rest. He lost 5 kg of his body weight during last 5 months. On rectal examination, fresh blood was seen around anal region and soft mass was felt on digital rectal examination. On proctoscopy, anal cavity and rectum were seen filled with fresh blood, but active site of bleeding, polyp, or ulcer was not detected. On laboratory examination, red blood cell count was $3.09 \times 10^{12}/L$ (Normal: $4.32-5.72 \times 10^{12}/L$) and hemoglobin was 86 g/L (Normal: 135–175 g/L). All other parameters were within normal limit. None enhanced CT showed isodense (35HU) homogenous bowel wall thickening that on contrast-enhanced CT venous phase enhances heterogeneously. Multiple calcifications were seen in the thickened bowel

wall and around the peri-rectal area. Lesion was seen extending from distal sigmoid to whole of the rectum (Fig. 1). Multiple hypodense lesions were also seen in spleen (Fig. 2).

After initial management of anemia, the patient underwent abdominal laparotomy followed by surgical excision. During surgery, 25 cm long lesion was found extending from distal sigmoid to whole of the rectum. Whole of the rectum and part of the sigmoid colon were excised and sigmoid-anus anastomosis was done. Postsurgical histopathological examination of excised specimen showed submucosal multiple thin-walled vessel of varying size with interposed stroma. Some vessels lumen consisted of blood cells (consistent with blood vessel), whereas other consisted of clear fluid (consistent with lymph vessel). Immunohistochemistry of specimen showed endothelial cells positive for CD 31 and CD 34. Some cells were positive for D2-40, while others were negative for D2-40 (Fig. 3). On the basis of histopathological report and immunohistochemistry, hemolymphangioma was diagnosed.

The surgery, which followed by complication (intestinal infection), was well managed and the patient was discharged

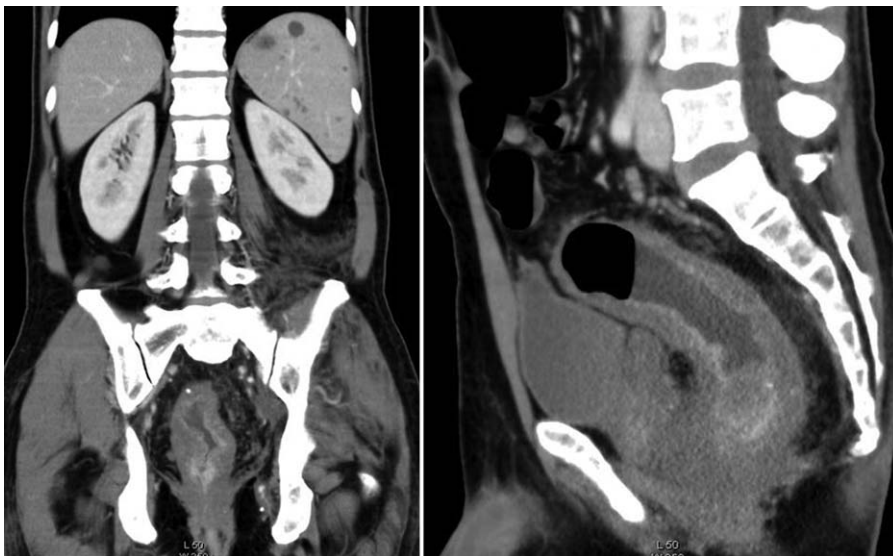


Figure 2. Coronal and sagittal view of abdomen. Coronal (left) and sagittal (right) view of abdomen, heterogeneously thickened bowel wall is seen extending from distal sigmoid to whole of rectum. On coronal image, multiple hypodense lesions are seen in spleen.

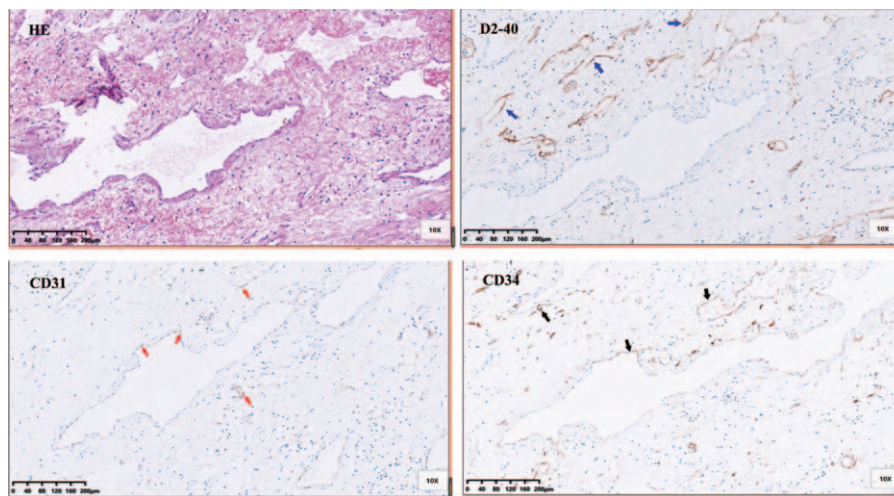


Figure 3. Histopathological examination of the rectal hemolymphangioma (hematoxylin and eosin staining). On H&E preparation, postsurgical histological specimen showed multilocular cystic structure covered with endothelial cells. Cyst wall is composed of dilated blood vessels, lymphatics, with inflammatory cell infiltration and abscess formation. Large lymphatic channels are seen positive for D2–40. Vascular endothelial showed positive for CD31 and CD34.

from hospital on the 23rd day of surgery. Then after, no further complication or recurrence was noticed during 6 months’ follow-up. This study was approved by the First Affiliated Hospital of Sun Yat-Sen University Institutional Review Board. Written consent for this case report was obtained from the patient.

3. Discussion

Hemolymphangioma is a rare developmental defect. The malformations result from disruptions in the development and maintenance of normal morphology and are believed to occur due to obstruction of veno-lymphatic communication between systemic circulation and dysembryoplastic vascular tissue.^[4,5] Venous malformations (VMs) can either be caused by somatic mutation or familial mutations, but lymphatic malformations (LMs) are caused by somatic mutations.^[6] Evidence for familial form of LMs is not yet detected. Thus, hemolymphangioma probably occurs due to somatic mutation in gene important for proliferation, migration, adhesion, and stability of the developing venous and lymphatic vasculature. The causative gene for both sporadic and familial VMs is believed to be due to mutation in the gene *TIE2/TEK* and endothelial cell-specific tyrosine kinase receptor.^[4] The cause for LMs is still unknown; some speculate the role of vascular endothelial growth factor C (VEGF-C) in lymphangiogenesis.^[7] The coexistence of these 2 entities in hemolymphangioma is not well understood, but it strengthens the hypothesis that both blood and lymph-endothelial cell originate from common stem cell.^[8]

Hemolymphangioma is believed to be embryological developmental error presented by birth. They can be localized or diffused and can occur as solitary or multiple lesions involving single or multiple sites. Our case had diffused hemolymphangioma in sigmoid colon and rectum along with suspected lesions in spleen. Deep and asymptomatic lesion can remain veiled for many years unless it is presented with symptoms or is diagnosed incidentally. Superficial skin lesion and lesion near to mucosa can be diagnosed early with bleeding complication in comparison to deep seated lesions. Clinically, the progression of disease can vary from stable or slow growing lesion to aggressive enlarging tumor without invasive potential.^[9] Although the exact modulator is not well established, hormonal factory, infection, inflammation,

and trauma can lead to rapid growth. Our case was a 57-year-old man, who first complained of rectal bleeding at the age of 2. After a long history of gradual increment in symptoms, he was presented with massive rectal bleeding since last 5 months. The factor affecting the rapid progression of disease in later days is not understood. He had history of multiple surgical interventions. Surgical trauma to the left over tissue can be one reason for rapid progression.

Radiologically, it is a great challenge to diagnose hemolymphangioma. Mix lesion of blood vessel and lymphatic vessel with interposed stroma in between give variable appearance according to the proportion of individual tissue. Lesion with cavernous lymphatic sac can appear multilocular cystic lesion with attenuation similar to water or corresponding serous and chyle. Dilated blood vessel can appear isointense with soft tissue, which on contrast examination shows higher attenuation in venous phase. Blood stasis in malformed and dilated blood vessel usually shows partial or complete thrombosis and is often associated with dystrophic calcification.^[2] Phlebolith is sensitive to VM, but false-positive results are possible in pelvic area where the dilated and torturous vein are common due to other cause. Spontaneous or traumatic rupture with hemorrhage/ lymphorrhea can cause inflammatory changes that chronically lead to fibrosis. Our case is a chronic case with multiple surgical interventions, so it showed nonspecific radiological finding. Bowel mucosa appeared thick and showed moderate enhancement on venous phase, which can probably be due to abundant malformed blood vessel in that region. Thus, this can also support the massive rectal bleeding. Other regions display heterogeneous enhancement, which can probably be due to mixed blood and lymphatic vessel with superimposed inflammation and fibrosis. Multiple phleboliths were seen in and around the lesion.

Besides diagnosis, radiological tests such as CT and magnetic resonance imaging (MRI) can also help find the extent of the lesion and plan the appropriate treatment. Nonsurgical method such as cryotherapy, laser therapy, radiotherapy, and local injection of sclerotic agents has been shown to be effective, but surgical excision is comparatively more superior.^[9] A thorough exploration of the abdominal cavity and complete excision are believed to provide the best result with lower recurrence rate.

In conclusion, hemolymphangioma of the rectum is a rare cause of rectal bleeding. It is believed to be embryological development error and all lesions are supposed to present by birth. Anatomical location and progression of disease determine the age of presentation. The lesion is a mixture of multiple tissues, thus radiological appearance is not specific and vary according to the content of the lesion. Therefore, radiological test alone can be a challenge in making diagnosis. Detail clinical history, physical examination, specific radiological findings, and follow-up can help make proper diagnosis and choose appropriate treatment.

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