

An Investigation of Clinical Features and Exploration of Comprehensive Treatments for Colorectal Vascular Malformation in Children

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Abstract

Objectives: To summarize the clinical features, treatment methods, and outcomes of the rare diseases of colorectal vascular malformation (CRVM).

Methods: We retrospectively analyzed clinical data of CRVM patients admitted to the Department of Pediatric Surgery of Children's Hospital of Fudan University from 2004 to 2019.

Results: A total of 23 cases (16 males, 7 females) were enrolled. The median age of symptom onset was 1.4 years. Eleven patients had been misdiagnosed, and the average time from symptom onset to diagnosis was 4.5 ± 4.4 years. Associated vascular malformations in the skin and other parts were found in eight patients. All patients showed a positive anomalous vascular image in the rectum and surrounding areas with contrast-enhanced computed tomography (CT) / magnetic resonance imaging (MRI). The sensitivity of endoscopy in the diagnosis of CRVM was 82.6%. A total of 21 patients underwent a modified Soave procedure. The lesions were mostly restricted to the rectum and sigmoid, with an average length of 20 ± 5.4 cm. The median follow-up time was 78 months. Bloody stools reappeared within 6 months in three patients, but were cured by sclerotherapy or fulguration. At the last time of follow-up, bloody stool symptoms were absent and hemoglobin was in the normal range for all patients.

Conclusions: CRVM occurs more commonly in male children than in female children. Combined with enhanced MRI, CT, and colonoscopy, CRVM diagnosis can be confirmed. The lesioned intestinal segment can be safely removed through a modified transabdominal Soave procedure, effectively curing or relieving hematochezia.

What Does This Paper Add To The Existing Literature?

The clinical features and diagnostic methods of a rare disease in children—colorectal vascular malformation (CRVM)—were analyzed in this paper. A transabdominal anal modified Soave procedure is introduced to remove the lesioned intestinal segment safely, and effectively cure or relieve hematochezia.

Introduction

Colorectal vascular malformation (CRVM) is a rare, benign angiogenic lesion. CRVM is most often located in the rectosigmoid region, with primarily venous malformations (formerly known as 'cavernous hemangioma') (1, 2). The most common clinical symptom is recurrent, painless rectal bleeding that presents with melena, hematochezia, and anemia. Due to the non-specificity of clinical presentation, CRVM is often misdiagnosed as hemorrhoids, colitis, polyps, or anorectal varices (1, 2, 3). Endoscopic treatment, angiographic embolization and surgical excision of the lesions have been reported to be effective in the treatment of CRVM (4, 5). However, due to the low incidence and unclear pathological mechanism, most of the previous studies were case reports, there is still a lack of consensus on diagnosis and treatment of CRVM in children. In this study, we reviewed 23 children with CRVM who were

treated in our center from 2004–2019 to summarize experiences related to the diagnosis and treatment of CRVM.

Patients And Methods

We reviewed data from 23 patients treated for CRVM in the Department of Pediatric Surgery of Children's Hospital of Fudan University from 2004 to 2019. Patients with concomitant hemangioma in the pelvic cavity, genitourinary tract, limbs, and skin were included. Hematochezia caused by simple hemorrhoid varices, portal hypertension, portal vein malformation, Abernethy malformation, inflammatory bowel disease, and other secondary colorectal varices or hyperplasia were excluded in this study. The clinical information that was collected included sex, age at symptom onset, age at diagnosis, severity of rectal bleeding, misdiagnosis, treatment process, clinical outcome, and lesion distribution range. We reviewed the consequence of assistant examinations, such as colonoscopy, CT, MRI, and histopathology. Follow-up was conducted through face-to-face interviews in the outpatient departments and also through telephone interviews. This study was approved by the Ethics Commission of Children's Hospital of Fudan University. This study was performed in accordance with relevant guidelines and regulations. The informed consent was exempted by the Ethics Commission of Children's Hospital of Fudan University.

Results

General situation

A total of 23 patients with CRVM were included in the study (16 males; 7 females). Basic patient information is listed in Table 1. The median age of bleeding onset was 1.4 years (range: 1 month–12 years) and most patients had been previously treated in hospitals. The median age at diagnosis was 6 years (range: 1–12 years) and the average time from symptom onset to diagnosis was 4.5 ± 4.4 years. Of the patients in the study, 23 (100%) had intermittent or persistent hematochezia and 20 patients (87%) had anemia. The mean patient hemoglobin level was 75.8 ± 27.8 g/l, and hemoglobin was lower than 90 g/l in 17 patients (74%). A total of 11 patients (47.8%) had history of repeated blood transfusion. Other symptoms included: mild lower abdominal pain in six patients, intermittent mild diarrhea in four patients, intermittent hematuria in two patients, varicose veins appearing at the anus during defecation in two patients, and occasional vaginal bleeding before puberty in one patient. Eight patients (34.8%) presented with vascular malformations in other locations. Two patients presented with skin vascular malformation of the trunk and extremities, accompanied by unilateral lower limb hypertrophy (Fig.1A). Other skin vascular malformations are distributed in the limbs, penis, buttocks (Fig.1B,C,D). Abnormal vascular mass in the bladder or urethra was observed in two patients (Fig.1E), and vaginal vascular malformation was observed in one patient.

Misdiagnosis and mistreatment

A history of misdiagnosis in local hospitals was identified in 14 patients(60.9%). Four patients were misdiagnosed with anal fissures, and three patients were misdiagnosed with internal hemorrhoids before undergoing hemorrhoidectomy. Two patients were misdiagnosed with rectal polyps, one of which underwent endoscopic polypectomy. Two patients were misdiagnosed with inflammatory bowel disease and were treated with anti-inflammatory and hormone therapies. Two patients were misdiagnosed with portal hypertension, and rectal varices were treated with varicose vein ligation under a colonoscope. One patient was misdiagnosed with Meckel's diverticulum and underwent laparoscopic exploration. Reduced bleeding was not achieved in any of the patients who received invasive treatment or conservative treatment.

Imaging studies

All 23 patients were examined by colonoscopy. Lesions were identified to have originated from the dentate line and mucosal lesions showed multiple focal distributions. The mean length of the involved bowel was found to be 14.3 ± 4.7 cm by colonoscopy. Abnormal submucosal vascular distribution could be seen by colonoscopy in 19 patients(82.6%; Fig.2), and typically presented during colonoscopy as segmented papular vascular plexus or reticular varicose vessels. The remaining four patients showed non-specific manifestations, such as mucosal hyperemia, swelling, and ischemia(Fig.2F,G,H). A contrast-enhanced CT or CT angiography(CTA) examination was performed in 17 patients(Fig.3). One or more positive signs, such as irregularly thickened rectal walls with varying degrees of enhancement, blurred perirectal spaces, or abnormal dilatation of blood vessels in or surrounding the rectum was found by CT scans in all tested patients. Eight patients(46%) were found to have multiple calcification foci representing phleboliths(Fig.3A). Meanwhile, CT analysis also showed some concomitant diseases. One patient had a cystic space occupying the anterior portion of the right iliac vessel, which was considered to be a lymphatic malformation. Contrast-enhanced MRI was performed in 10 patients, which typically showed focal or multiple annular thickening of the rectal wall with low or equal signal intensity on T1 weighted image(T1WI), as well as significantly increased signal intensity of T2WI and fat suppressed(FS)-T2WI (Fig.3D,E,F). Moreover, MRI revealed the involvement of the pelvic cavity in two patients and the buttocks in another patient.

Treatment and Follow-up

A total of 21 patients underwent a modified Soave procedure to remove the diseased intestinal segment; Sarasola-Klose hemorrhoidectomy was used for the distal portion of the endorectal dissection(6). Three of these patients underwent a laparoscopy-assisted Soave procedure. Simultaneous loop ileostomy was performed in 18 patients; enterostomy was closed after a colonoscopy examination 3–6 months later. One patient underwent a redo pull-through transection and anastomosis because of intestinal necrosis and anastomotic stricture. Two patients with bladder vascular malformations who presented with focal eminence and erythema were subsequently cured by cystoscope electrocauterization. Urethral vascular malformations were not treated, and there was no recurrence of hematuria in the follow-up. The patient with massive vaginal bleeding achieved a significant remission after the Soave procedure, which may

benefit from suture and ligation of the abnormal hyperplastic blood vessels around the uterus as much as is possible. One patient diagnosed with KTS presented with occasional, slight rectal bleeding and was treated with conservative management and iron supplements. One patient with limited lesions underwent endoscopic foam sclerotherapy every 6 – 12 months.

Five of the 23 patients failed to follow-up after surgery; the other 18 patients were followed for 6–184 months after surgery (mean: 78 months). Bloody stools reappeared within 6 months in three patients, and colonoscopy showed abnormal angiogenesis near the muscular rectal cuff in two patients; bloody stool disappeared after electrocautery or sclerotherapy. One patient who received endoscopic sclerotherapy was injected with lauromacrogol again because of hematochezia recurrence. Two patients showed an increase in defecation frequency after enterostomy closure, and were remitted with 6 months of conservative treatment. In three patients, intermittent fecal soiling was found within 3 months after the enterostomy closure. Two of these patients recovered gradually 3–8 months after surgery. All children had no bloody stool symptoms, and hemoglobin was in the normal range at the last time of follow-up.

Distribution of the CRVM

The lesions of 21 patients showed segmental diffuse distribution. The boundary of the normal intestine was clear, combined with the results of intraoperative exploration. Vascular tortuosity and dilatation could be seen on the serosa of the intestinal wall, and thickened and twisted blood vessels existed in some of the mesenteric vessels (Fig. 4A,B,C). The lesions were mainly involved in the rectum, sigmoid colon, and descending colon; among them, two patients showed jumping distribution. In addition to rectal sigmoid colon involvement, ileum and transverse colon were also involved. Four patients were found to have anal canal involvement (Fig. 4D). The average length of the involved intestine was 20 ± 5.3 cm, which was significantly longer than that of the lesions seen by colonoscopy (14.3 ± 4.7 cm $P = 0.0158$). Upon histopathological examination, vascular malformations were shown to involve the whole intestinal wall in 11 patients, the submucosa and muscle layer in seven patients, and the submucosa in three patients. Histopathology confirmed that primary vascular malformations were venous in nature, and a few of them were complicated with lymphatic malformations (Fig. 4E).

Discussion

CRVM is a rare, gastrointestinal vascular malformation. Since CRVM was first described by Phillips 1839, more than 300 cases have been reported, and most of these have been in adults (7). Approximately 80% of CRVM lesions are located in the rectosigmoid (1, 2). The vascular malformation can present as solitary lesions, clusters of intraluminal polypoid lesions, or diffuse infiltration in the submucosa and the other layers of the intestinal wall with extension into the mesentery and adjacent organs (8). The classification and terminology of hemangioma and vascular malformation mentioned in the literature are not entirely clear, and mainly include cavernous hemangioma, angiodysplasia, arteriovenous malformation (AMV), venous malformation, and venous dilatation (9, 10). At present, CRVM is primarily reported to be a venous malformation (formerly called 'cavernous hemangioma'), and arteriovenous malformations are rare (6, 5,

10). Previous studies have reported that CRVM affects more adult females than adult males(11), but more male children than female children(12). In our study, the results showed the morbidity of CRVM in male children was 69.6%(16/23), which is consistent with previous reports.

Chronic, recurrent, painless rectal bleeding is the most common symptom of CRVM. Occasionally, massive and life-threatening bleeding may occur(1, 6). Most patients often have a long-term and history of repeated blood transfusions throughout the course of the disease. Other reported, but rare, symptoms include abdominal discomfort, abdominal pain, diarrhea, and perianal discomfort(2, 8). CRVM may manifest as intestinal lesions in syndromic diseases, such as KTS, which is characterized by CRVM, skin capillary malformation, and asymmetric limb hypertrophy(13, 14). In this study, two children showed multiple wine-like skin spots all over the body, complicated with asymmetric hypertrophy of both lower limbs, which was considered to be KTS. Due to the rareness of CRVM and the lack of specific symptoms, a substantial number of patients have been misdiagnosed with anal fissure, hemorrhoids, rectal polyps, inflammatory bowel disease, and rectal varices. Wang et al. reported that the mean delay time between the initial symptom onset and final CRVM diagnosis was 17.63 years(2). However, the onset of CRVM in more than 50% of adults can be traced back to childhood or even infancy(1, 2). Therefore, physicians should carefully consider the possibility of CRVM in children who complain of rectal bleeding.

Due to the low incidence of CRVM and the lack of large sample data, no consensus has been formed with respect to diagnostic methods. Colonoscopy is believed to be the most useful visual examination as it depicts the luminal extension and morphology of the malformation(1, 2, 8). However, if the examiner is not experienced enough, atypical presentation by colonoscopy may lead to misdiagnosis. In the present series, 19 patients (82.6%) showed abnormal submucosal vessels with multiple focal, diffuse distributions by colonoscopy. The other four patients showed atypical mucosal or submucosal lesions, such as hyperemia, swelling, scar-like changes, and multiple thrombosis. We speculate these features seen at initial colonoscopy may have led to a diagnosis of inflammatory bowel disease.

Contrast-enhanced CT is another important examination tool to confirm a CRVM. The representative findings of CT include an irregularly enhanced and thickened rectal wall or perirectal tissue, and multiple calcified foci of pelvic phleboliths(1, 2). Some of the CT findings of CRVM are nonspecific. However, when combined with CT angiography(CTA), diagnostic efficiency can be greatly improved. CTA can clearly show even the smallest vessels of the malformation, as well as connections with other vessels of the circulatory system(15). This has important guiding significance for intraoperative bleeding control. MRI has better tissue resolution than CT, which is of great significance in evaluating the extent of intestinal wall invasion and surrounding organ involvement, and is important for guiding the choice of treatment(15). Furthermore, dynamic contrast-enhanced MR angiography(MRA) can be used to differentiate between high- and low-flow lesions(16). Typical imaging MRI findings show the rectosigmoid wall was markedly thickened with a high signal intensity on T2WI, an enhanced dynamic scan showed progressive enhancement. The addition of fat suppression on a fast spin-echo T2WI sequence is beneficial in depicting the true extent of the lesion, as it helps to differentiate the hyperintense lesion from the rectal mesentery. Digital subtraction angiography(DSA) can help to visualize

the lesion and identify a vessel for embolization(4), but had a low positive rate of rectal vascular diagnosis(5/13, 38.5%) and could not show the lesion range(2). With its low sensitivity, invasiveness, and high price, DSA can be replaced by CTA and MRA. Therefore, we suggest that enhanced CT/CTA and/or MRI/MRA are recommended as first-line examination methods for children with typical CRVM clinical manifestations.

Endoscopic ultrasonography has a good application prospect in the diagnosis of CRVM, because it can accurately determine the level of intestinal wall involvement(8, 17). It can also evaluate the depth and direction of hemangiomas in the lower rectum and anal canal, especially for vascular abnormalities involving only the plasma layer, which cannot be recognized by colonoscopy. However, endoscopic ultrasonography has not been widely carried out in most children's endoscopic centers in China, and therefore, its application is limited.

CRVM treatments reported in the literature include surgical resection of the involved intestines, interventional embolization, and endoscopic treatment(1, 4, 18). Surgical resection of the diseased intestinal segment is one of the more effective methods for treating CRVM. However, there is yet to be a consensus regarding specific surgical methods. The earliest reported surgical treatment was abdominoperineal resection(APR), which involves removal of a diseased intestinal segment to effectively stop bleeding(2). Although this approach can save some patients with life-threatening severe intestinal bleeding, a permanent colostomy is undesirable, especially in young patients. Low anterior resection(LAR) can ultimately preserve sensation and defecation function of the anus(19); however, LAR cannot completely remove the diseased rectum and the probability of recurrent bleeding is very high. Therefore, this method can only be used for the treatment of vascular malformations above the sigmoid colon. In 1976, Jeffery first advocated for coloanal sleeve anastomosis for the treatment of CRVM, and this procedure has been remained popular since(20). However, this method is only able to remove the mucous layer and submucosa of the colorectum and retain a 10-cm-long rectal seromuscular layer. Therefore, vascular malformation of the seromuscular layer may invade the colon, and cause bleeding recurrence. It is also very difficult to peel off 10 cm of mucous membrane and seromuscular layer in the rectal wall with vascular malformation.

Recently, transanal total mesorectum excision(TaTME) for diffuse cavernous hemangioma of the rectum has been reported to contribute to the precise resection of the local lesion in the mid and low rectum and the anal canal with satisfactory outcomes, when applied in rectal cancer patients(1, 21). However, this operation completely removes the internal rectal sphincter and perirectal nerves, significantly influencing defecation sensation and defecation reflex functions after the operation. There is also a high risk of anastomotic leakage between a full-layer colon and low anal anastomosis.

Previous reports on surgical methods are mostly limited to adult cases. Due to the particularity of the anatomical structure of children, the adult procedure cannot be used entirely for reference. Previously, we used a modified transabdominal Soave procedure combined with Sarasola-Klose internal hemorrhoidectomy to treat CRVM, and achieved satisfactory results(6). In this procedure, the laparotomy

or laparoscopy was used to explore the extent of intestinal, mesenteric, and pelvic vascular malformations. The diseased mesentery is disconnected, and the colon and rectum are completely disintegrated below the peritoneal reflection, 3–5cm proximal to the dentate line. The extent of disassociation can be determined by digital rectal examination during the operation. Turn to the perineum, a gauze roll of 1.5 cm in diameter is slowly inserted into the rectum to a depth of about 3–5 cm. The distal end of the gauze roll and the rectal mucosa proximal to the dentate line are sutured. Circumferential incisions are made on the rectal mucosa's dentate line, and the gauze coils are pulled outward at the same time. Along the submucosal plane, the dissection is continued for another 3–5cm proximal to the dentate line and enters the abdominal cavity. Then, the bowel is pulled down until normal bowel appears. After the diseased bowel is resected, the cut edges of the colon are anastomosed to the dentate line. The internal sphincter and rectum muscle sheath are preserved, and the dissection range of the rectal mucosa is shortened in this procedure. Thus, the difficulty of operation and the risk of bleeding are reduced greatly. A gauze roll is used to assist in achieving hemostasis by direct compression during dissection of the mucosa. The mucosa is fixed and retracted with tension, allowing the incision to be regular and the dissection plane to be well controlled. Due to the short distance from the peritoneal reflection to the dentate line in children, preserving a 3–5cm rectal muscle sheath can significantly reduce perirectal nerve injury and minimize the effects on defecation.

In our center, 21 CRVM patients were treated with modified Soave surgery for complete resection of the diseased intestinal segment. Only two patients presented with abnormal vascular hyperplasia near the anastomotic site and hematochezia; these were cured by endoscopic electrocautery. There was no reoperation due to the recurrence of hemangioma caused by the preservation of the muscle sheath. Therefore, even if the lesion begins from the dentate line, minor vascular malformations that might be retained in the bowel wall distal to the anastomosis after these resections can be easily managed by conservative treatment. Some children have a poor sense of defecation after operation, likely due to the removal of the sensitive area of the mucous membrane on the dentate line, which requires slow recovery. Similar to the endorectal pull-through in the procedure for Hirschsprung's disease(22), we noted an increased stool frequency in the early postoperative stage. This returned to normal by several months after the operation. Therefore, the modified transabdominal transanal Soave procedure is a good method for treatment of CRVM because it can significantly reduce operation difficulty, bleeding risk, and postoperative defecation dysfunction.

In addition, our study found that mucosal or submucosal lesions of the intestinal wall identified by colonoscopy were significantly smaller than that of serous side lesions seen during operation. Children with long-segment lesions often showed patchy or small segment lesions by colonoscopy. Therefore, transabdominal surgery to explore the extent of the lesion is of great significance to prevent the omission of the diseased intestine and to prevent postoperative rebleeding. Pelvic floor vascular malformation around the peritoneal reflection can be seen during the abdominal exploration in some children, which requires ligation around the rectum to control bleeding. In children with uterine and vaginal bleeding, the primary abnormal blood vessels around the uterus can be ligated during the operation to reduce vaginal

or menstrual bleeding. Moreover, for a small number of children, abdominal lymphatic vascular malformation can be excised simultaneously with transabdominal exploration.

Nonoperative therapies to treat CRVM, such as sclerotherapy, snare polypectomy, neodymium-doped yttrium aluminum garnet (Nd:YAG) laser therapy, and argon plasma coagulation (APC), have been reported to decrease the amount and frequency of hematochezia(23-25). However, rectal bleeding eventually recurred in most patients. In our center, one patient with limited lesions was treated with sclerotherapy and had recurrent hematochezia. The scope of rectal submucosal vascular malformation was gradually increased during follow-up. Moreover, sclerosing agent injection and interventional vascular embolization increased the risk of deep venous thrombosis, intestinal necrosis, pulmonary embolism, and even death. Data from several studies suggest that medical therapies is an effective and relatively safe treatment for patients with refractory bleeding from gastrointestinal vascular malformations(11, 26). Smith et al. reported that the patient was successfully managed using a conservative approach with tranexamic acid administered as needed, avoiding the need for resection(7). However, the long-term efficacy of drugs in the treatment of CRVM remains inconclusive.

Declarations

Yanbing Huang and Menghua Xu contributed equally to this work as first authors.

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Tables

Table 1. Clinical Data of 23 CRVM Cases	
Variables	N=23
Gender, n (%)	
Male	16(69.6)
Female	7(30.4)
Age (y)	
Mean ± SD	2.8
Median (range)	1.4(0.1-12)
Time to diagnosis (y)	
Mean±SD	7.1
Median (range)	6(1-12)
Symptoms, n (%)	
Hematochezia	23(100)
Anemia	20(87)
Abdominal pain	6(26)
Other vascular malformation	8(34.8)
hematuria/Vaginal bleeding	3(13)
Hemoglobin (g/dL)	
Mean±SD	75.8
Median (range)	67(39-120)
Blood transfusion, n (%)	
Yes	11(47.8)
No	12(52.2)
Misdiagnosis ,n(%)	
Anal fissure	4(17)
Hemorrhoids	3(13)
Colitis	2(8.7)
Varicosis	2(8.7)
Polypus	2(8.7)

Location, n (%)	
rectum	23(100)
Sigmoid colon	20(66.7)
anal canal	4(17)
SD = standard deviation.	

Figures



Figure 1

A Asymmetry of lower extremities with the right leg demonstrating soft tissue and bony hypertrophy, as is typically seen in KTS. B, C Cutaneous vascular malformation of the lower limbs and perineum. D Port-wine stains on the buttocks. E abnormal vascular mass in the bladder triangle.

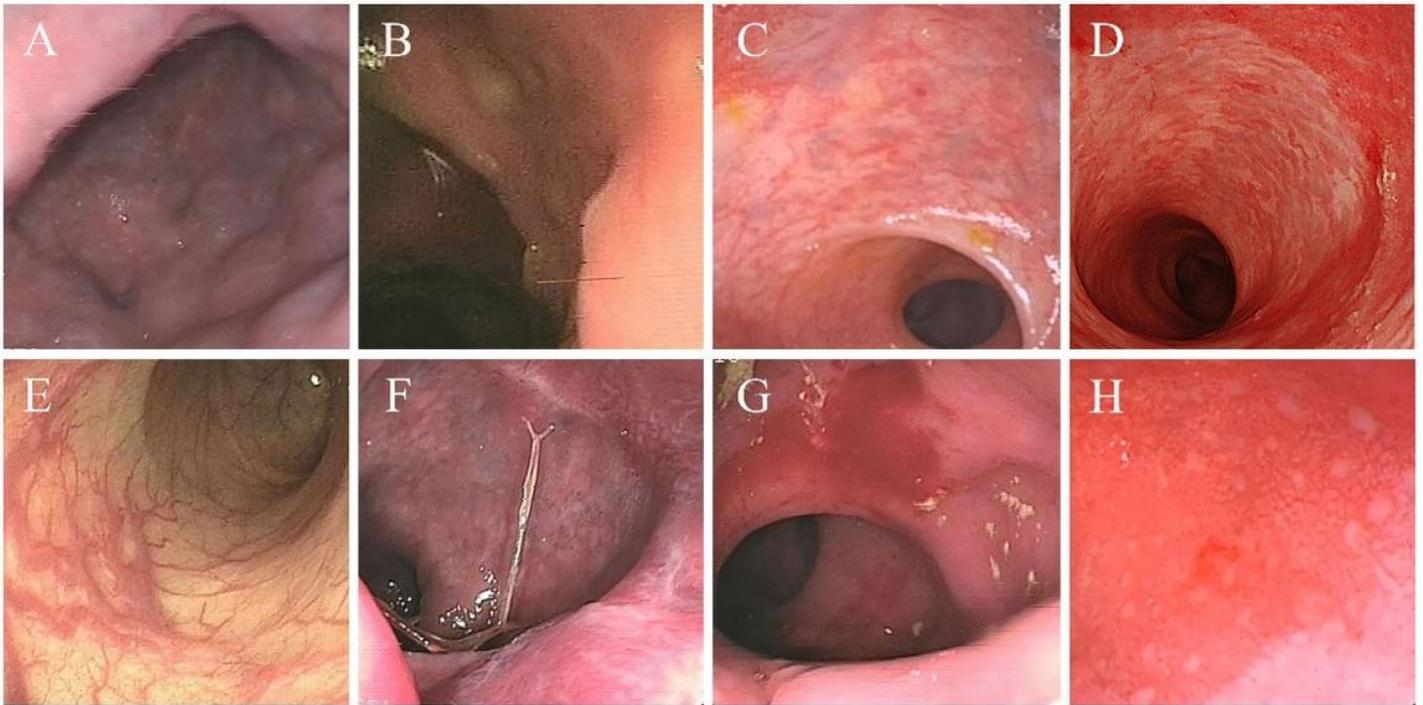


Figure 2

Colonoscopic findings of CRVM. A Serpentine dilated vessel. B Blue-purple nodules. C Sporadic blue patchy changes. D Mucosa ranging from bright red to bluish-purple. E Vascular thickening with prominent exposure. F Ischemic manifestations (scar-like changes). G Congestion, swelling, and bleeding. H White, round, spot-like changes.

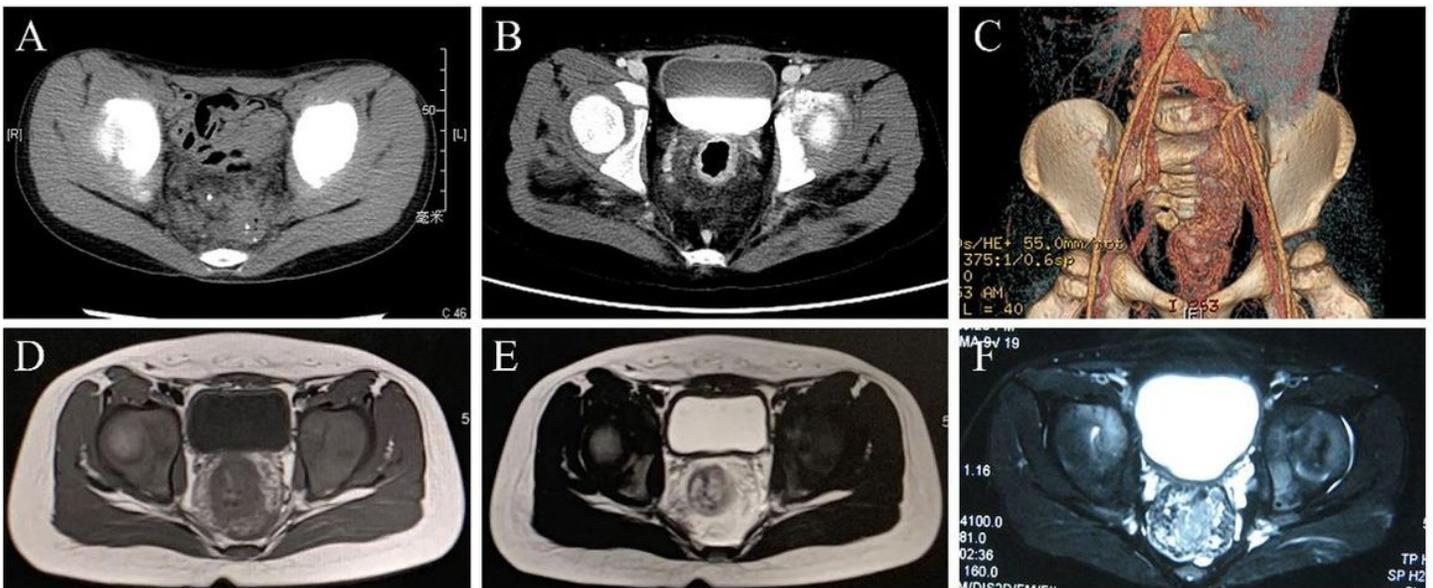


Figure 3

CT and MRI findings of CRVM. A Thickened rectal wall and multiple calcifications. B Circular enhancement of rectal mucosa. C CTA shows more collateral branching of the venule around the rectum. D T1-weighted axial. E T2-weighted axial. The lesions were hypointense on T1WI and significantly hyperintense on T2WI. F A large number of tortuous cord-like signals in the mesorectum on the FS-T2WI.

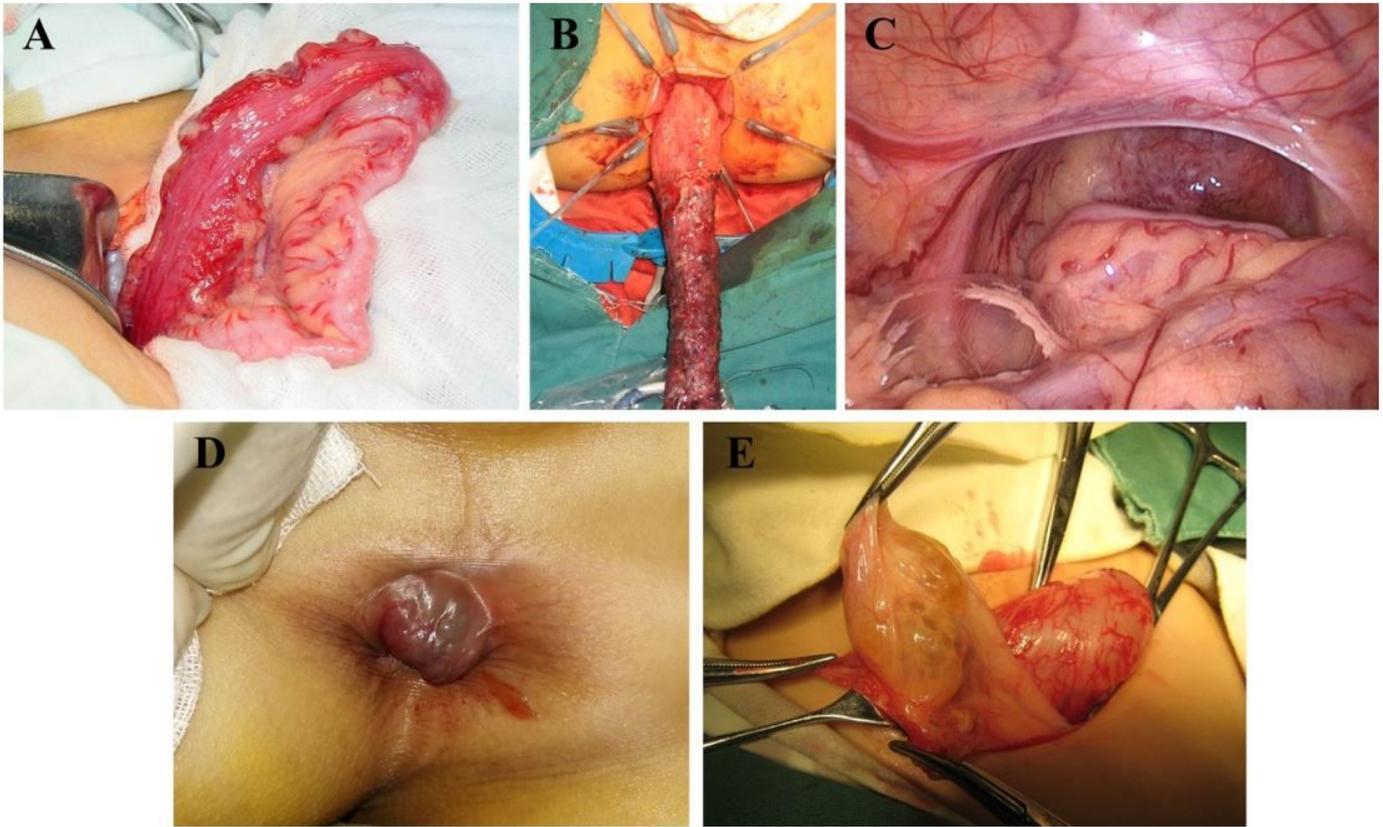


Figure 4

A Tortuous and enlarged blood vessels on the surface of the intestine. B Vascular malformation was noted in the rectum and sigmoid colon. C Distribution of abnormal blood vessels in the pelvic cavity. D Vascular malformation involving the perianal region. E Lymphatic malformation.