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Case report

Colonic polyposis in a 15 year-old boy: Challenges and lessons from a rural resource-poor area

Nasser Kakembo [a], Phyllis Kisa [a], Tamara Fitzgerald [b], Doruk Ozgediz [c, *], John Sekabira [a]

[a] Department of Surgery, College of Health Sciences Makerere University, Uganda
[b] Texas Tech University, USA
[c] Yale University, USA

HIGHLIGHTS

- Polyposis syndromes present unique diagnostic and treatment challenges in resource-limited settings.
- Presentation may be with chronic symptoms and advanced disease.
- History and physical exam provide valuable information when other imaging and diagnostic modalities are not available.
- Previous reports from similar settings describe partial colectomies.
- Total proctocolectomy with ileal pouch reconstruction is feasible and safe.

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ABSTRACT

Introduction: Colorectal polyps usually present with rectal bleeding and are associated with increased risk of colorectal carcinoma. Evaluation and management in resource-poor areas present unique challenges.

Presentation of case: This 15 year-old boy presented with 9 years of painless rectal bleeding and 2 years of a prolapsing rectal mass after passing stool. He had 3 nephews with similar symptoms. On clinical assessment and initial exam under anesthesia, an impression of a polyposis syndrome was made and a biopsy taken from the mass that revealed inflammatory polyps with no dysplasia. He was identified during a pediatric surgical outreach to a rural area with no endoscopy, limited surgical services, and no genetic testing available, even at a tertiary center. He subsequently had a three-stage proctocolectomy and ileal pouch anal anastomosis with good outcome after referral to a tertiary care center. The surgical specimen showed many polyps scattered through the colon.

Discussion: In the absence of endoscopic surveillance and diagnostic services including advanced pathology and genetic testing, colorectal polyposis syndromes are a significant challenge if encountered in these settings. Reports from similar settings have not included this surgical treatment, often opting for partial colectomy. Nonetheless, good outcomes can be achieved even given these constraints. The case also illustrates the complexity of untreated chronic pediatric surgical disease in rural resource-poor areas with limited health care access.

Conclusion: Polyposis syndromes in children present unique challenges in rural resource-poor settings. Good outcomes can be achieved with total proctocolectomy and ileal pouch anastomosis.

1. Introduction

Colorectal polyps commonly present with rectal bleeding in children, adolescents and adults. Multiple juvenile polyps pose an increased risk of intestinal cancer if there are more than 5 [1]. Most children with juvenile polyps have 1–5 polyps, and genetic syndromes can predispose some children to adenomatous polyposis and hamartomatous polyps. The presence of multiple adenomatous polyps in the large bowel confers a 100% lifetime risk of colorectal cancer and generally mandates total proctocolectomy in the first or second decade of life, and mandates colonoscopic surveillance for

* Corresponding author. Department of Surgery, Yale University, USA.
E-mail address: dozgediz@hotmail.com (D. Ozgediz).

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neoplasia [2,3]. Although the presence of multiple adenomatous polyps (>100 polyps) can be due to mutations in the adenomatous polyposis coli (APC) gene, a large group of patients remain with multiple (5–100) adenomas and in whom there is no detectable APC mutation [2,4].

The hereditary polyposis syndromes include adenomatous polyposis syndromes (Familial Adenomatous Polyposis, Gardner syndrome) and the hamartomatous polyposis syndromes (Peutz-Jeghers syndrome, Juvenile polyposis, and Cowden's disease) [5]. The adenomatous polyposis syndromes are characterized by numerous adenomatous polyps throughout the entire colon and a spectrum of extra-colonic manifestations. They invariably progress to colorectal cancer without appropriate intervention [2,5]. Diagnosis of symptomatic polyposis is by colonoscopy, and here we present a teenager with colorectal polyposis from a resource-limited setting where there were no pediatric colonoscopy services posing diagnostic and treatment challenges.

2. Presentation of case

During a free pediatric surgical outreach camp to rural Uganda, a 15 year-old boy presented for screening with 9 years of rectal bleeding and 2 years of a red mass protruding from the anus after passing stool. The mother noted that he was staining the family pit latrine with fresh blood whenever he would pass stool, and for many years he had been treated with herbs from traditional healers with no improvement. Two years prior to presentation he noticed a grape-like red mass protruding through the anus that was associated with pain and fresh bleeding on defecation. Initially the mass would spontaneously reduce, but later it would remain prolapsed. He would sometimes become very pale and suffered from generalized body weakness and his school attendance remained very limited. He had not taken any formal medication and he had no history of surgery or blood transfusion. He was from a peasant family with no first-degree relatives with a similar problem but he reported three nephews who have occasional rectal bleeding. The boy was stunted compared to other siblings and children in the same community.

On examination he was moderately pale with no jaundice and no edema. He was generally of short stature and weighed approximately 35 kg. Abdominal exam was normal, and oral exam was normal with no hyperpigmentation of the lips. Digital rectal examination revealed a normal perianal area and sphincter tone. An exam under anesthesia was done at presentation and revealed a large boggy fungating mass in the rectum about 8 cm proximal to anal verge, consistent with a rectum matted with polyps. They were friable, circumferential, and easily prolapsed out of the anal canal with significant bleeding (Fig. 1). A clinical impression of possible adenomatous familial colorectal polyposis was made, and a biopsy was taken from the mass. The biopsy was subsequently processed in the capital city of Kampala and revealed inflammatory polyps without evidence of dysplasia.

Given the presence of a large number of rectal polyps, ideally, endoscopy and colonoscopy would have been performed to more fully assess the upper and lower GI tract, however, these services were not available. In addition, genetic testing was not available. We counseled the family of the concern for ongoing bleeding and longer-term cancer risk associated with polyposis syndromes, and offered 3-stage proctocolectomy with J-pouch reconstruction at a tertiary center, to which they consented.

At initial laparotomy several weeks later at the tertiary care center, the peritoneal cavity was clean and liver appeared normal with no tumor seeding. He had multiple enlarged lymph nodes at the ileocecal junction. The entire colon was inspected and palpated for polyps from cecum to rectum and multiple boggy masses were felt in the cecum and sigmoid colon. A small colotomy was done in the cecum revealing multiple polyps. Total abdominal colectomy was done excluding proctectomy, leaving the child with a temporary end ileostomy. The specimen was cleaned and opened lengthwise, grossly showing multiple pedunculated polyps (over 8 polyps) in the cecal area and descending colon (Figs. 2 and 3). There also appeared grossly to be numerous sessile polyps in the cecum and descending colon. The transverse colon appeared to be free of polyps. Pathology revealed inflammatory polyps with no evidence of dysplasia, and the lymph nodes in the specimen were benign. He tolerated the ileostomy well without dehydration, and the family was counseled about fluid intake especially given their rural village home environment.

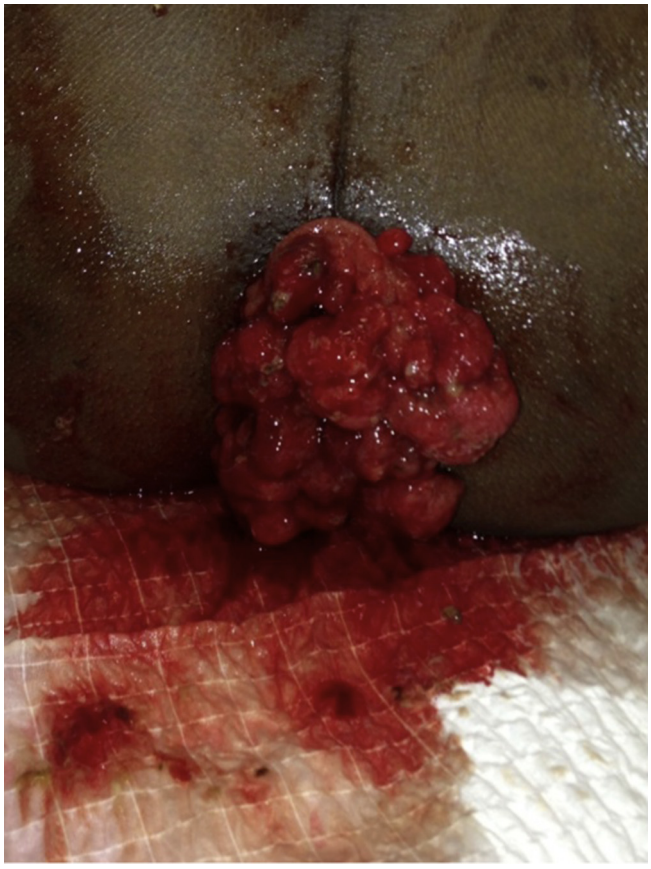

Fig. 1. Prolapsing rectal mass with innumerable polyps.

He returned 3 months later for proctectomy and J pouch ileoanal anastomosis. A 7 cm ileal J pouch was constructed in a stapled fashion using a donated stapling device, and a handsewn ileoanal J pouch anastomosis was constructed, with a protective diverting loop ileostomy. He tolerated the procedure well and once again recovered at home with the ileostomy. The final pathology of the proctectomy also was consistent with innumerable inflammatory polyps with no dysplasia. He returned several months later for ileostomy takedown and is doing well at one year of follow up. He has 4–5 bowel movements a day with no incontinence or leakage and has a normal rectal exam with no stricture or masses. He has returned to school and gained weight. We have recommended annual rectal exams, as possible, to monitor the several millimeters of very distal mucosa. The family was advised that the affected relatives should also present for surgical evaluation.

In preparation of this report, the CARE criteria were followed as referenced [6].

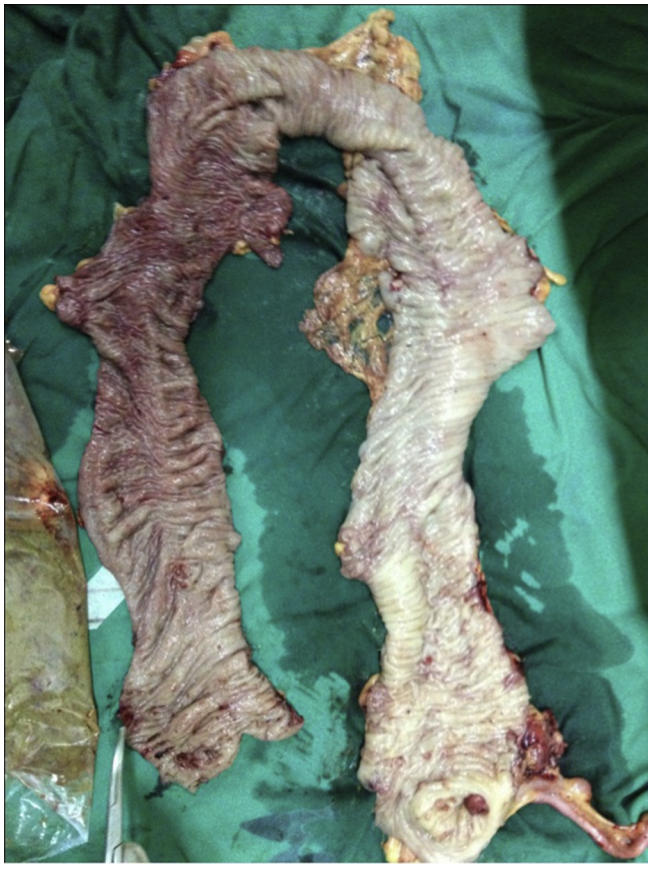

Fig. 2. Colectomy specimen. Note multiple polyps in cecum and descending colon.

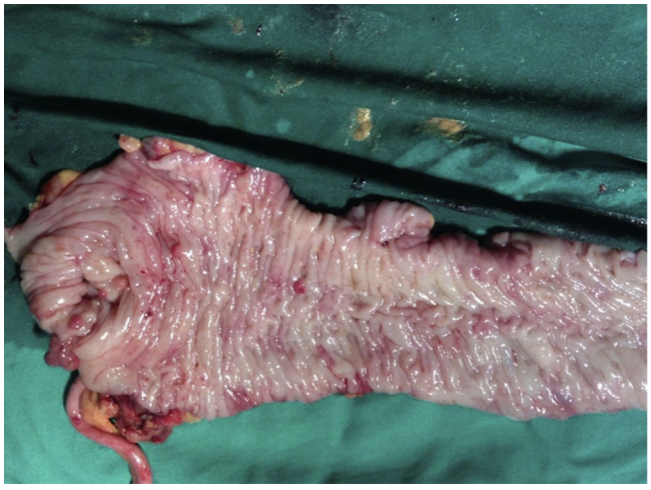

Fig. 3. Closer view of cecum, note multiple polyps especially concentrated at ileocecal junction.

3. Discussion

Colonic polyps usually present with rectal bleeding, but may also present with a prolapsing rectal mass, abdominal pain, mucopurulent discharge, diarrhea, and vomiting. Some patients are asymptomatic [7]. Less common forms of presentation include intussusception and auto-amputation of the polyps [8]. This boy presented with excessive bleeding after stooling with a prolapsing rectal mass. On presentation, our patient had innumerable polyps in the rectum, but the more proximal colon could not be assessed.

The World Health Organization criteria for diagnosis of juvenile polyposis syndrome are either more than five polyps in the colon or rectum, polyps throughout the gastrointestinal tract, or any number of juvenile polyps in a person with a family history of juvenile polyposis [2,9]. Our patient fulfilled all the criteria. Patients at risk or with high suspicion of juvenile polyposis should have endoscopic screening of the colon and upper gastrointestinal tract at 15 years of age or at the time of onset of first symptoms [2]. The surgical options for symptomatic juvenile polyposis syndrome are subtotal colectomy with ileorectal anastomosis, or total proctocolectomy with J pouch reconstruction [2,5]. In view of the risk of rectal cancer or intractable proctitis, ileal pouch anal anastomosis may be a safer alternative, if technically feasible, in the long-term in spite of possible inferior bowel function and increased frequency of stools [10]. Patients who undergo subtotal colectomy require routine endoscopic surveillance of the remaining rectum every 6 months for recurrent polyps or carcinoma, which requires access to endoscopy services that are not usually available in resource-poor areas [11].

There are very limited reports of such cases in resource-poor settings. A similar case reported from an Ethiopian tertiary care center in a boy who presented with a prolapsing rectal mass was treated with total abdominal colectomy and ileorectal anastomosis, proctoscopic diathermy, and plans for proctoscopic surveillance [12]. In a recent Nigerian case series, where colonoscopy was not available, digital rectal exam and transanal polypectomy was done in 16 children up to 7 cm from the anal verge, with no recurrent bleeding but unspecified median follow up for these patients. No evaluation could be performed of the more proximal colon [13]. The rectum in these cases, however, was not carpeted with polyps, as in our case. A similar report from Pakistan stressed that the majority of juvenile polyps could be identified on digital exam and sigmoidoscopy, and they did not perform more proximal endoscopy, though it was not specified whether this was due to resource limitations [14].

In a series of 17 patients from India, where colonoscopy was available, the majority of patients were treated with colonoscopic polypectomy, while six patients who could not have their polyps cleared went on to have total colectomy, mucosal proctectomy, and straight ileoanal anastomosis. Those who had colectomy and were followed up over several years settled to an average of seven bowel movements daily [15]. A report from Thailand of 32 patients emphasized that most polyps were in the rectosigmoid, but 20% of patients had right-sided polyps, as in our patient [16]. Surgical management was not discussed in this report. The largest North American series (of 36 patients) emphasized low malignancy risk in childhood and possibly longer intervals for colonoscopic surveillance [17].

We did not identify any reports of patients from a similar resource-limited setting treated with an ileoanal J-pouch anastomosis as definitive surgical therapy. Our patient's operations were staged due to his anemia, poor nutritional status (assessed clinically) at presentation, the need to evaluate the colon, and to evaluate for any gross sign of metastatic disease. We had concerns about dehydration from temporary ileostomy given the rural setting, but he was able to keep up with oral intake, and loperamide was not used.

As is the case for many chronic surgical conditions in rural areas, his presentation was greatly delayed after the onset of symptoms and led to years of morbidity, manifest as poor nutrition, bleeding, pain, social exclusion, and limited school attendance. This type of scenario is typical for chronic untreated surgical conditions in children. Barriers in access to care are multifold, and often include

poverty, poor education, limited trust in the local health care system, cultural beliefs, and fear, among many others [18]. This type of case encountered in rural surgical camps also suggests a likely significant underdiagnosis and treatment of these types of diseases in limited-resource areas.

In summary, a review of the key learning points and added knowledge from this case include: 1) polyposis syndromes present unique diagnostic and treatment challenges in resource-poor areas with patients often presenting with advanced disease; 2) in many such settings endoscopy may not be available and the history and physical examination are critical; 3) there are limited reports from similar settings, and the surgical management has mostly been partial colectomy; 4) our case shows that a j pouch ileoanal reconstruction can be performed with good outcome even in resource-limited areas and provides a long-term surgical solution with reduced need for ongoing surveillance.

4. Conclusion

Lack of colonoscopy services pose significant challenges for children with various gastrointestinal disorders in resource-poor areas. Nonetheless, digital rectal exam can yield valuable information. This patient had years of symptoms prior to presentation to health services, a situation that is also typical in resource-poor areas. Rural pediatric surgical outreach programs may identify such cases amenable to treatment in a tertiary center, and this was one of the objectives of our program. The outreach program allowed for the family to interact with formal medical services and facilitated a history and a brief exam under anesthesia. Clinically, the child had significant rectal prolapse at presentation with innumerable rectal polyps. Given the lack of endoscopy services, risk of ongoing bleeding, and long-term risk of cancer, we opted for total proctocolectomy with J pouch reconstruction in 3 stages. On gross and histologic examination of the colon once removed, the overall diagnosis was most consistent with juvenile polyposis. He tolerated the procedures well. The procedure has not been previously reported in a similar context and may be an alternative if technically feasible in such cases.

Ethical approval

Consent was granted from the patients family for the case report.

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None.

Author contribution

NK, PK, DO, JS, TF: all contributed to taking care of this patient, study design, data collection, analysis, and writing.

Conflicts of interest

None.

Guarantor

Doruk Ozgediz.
Nasser Kakembo.
John Sekabira.

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