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CASE REPORT | COLON

Cap Polyposis: An Elusive Diagnosis in a Pediatric Patient Successfully Managed With Endoscopic Treatment

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ABSTRACT

Cap polyposis is a rare condition of the rectum or sigmoid colon manifested by inflammatory polyps covered by a thick layer of fibrinopurulent mucus. This condition typically presents as mucoid diarrhea and rectal bleeding, and patients are often prescribed antibiotics (such as those for *Helicobacter pylori*), steroids, infliximab, or aminosalicylates. Surgical management is an option for unresponsive disease, but endoscopic management has been rarely reported. For cases of cap polyposis in which conservative medical management fails, wide-field endoscopic mucosal resection is a viable option.

INTRODUCTION

Cap polyposis is a rare benign disease of the colon, primarily the lower rectum, in which inflammatory polyps are capped by a thick layer of fibrinopurulent mucus and inflammatory granulation tissue. Since it was first described in 1985 by Williams et al,[1] fewer than 100 cases of cap polyposis have been reported.[2] Most cases have involved adults of ~50 years presenting with constipation, mucous discharge, diarrhea, abdominal pain, and rectal bleeding.[3] The polyps in this disorder can vary in size, shape, number, and location. In pediatric cases, polyps are typically confined to the rectum and rarely with extension to the anal canal or sigmoid colon.[2] The differential diagnoses include inflammatory bowel disease and severe pseudomembranous colitis/proctitis.[4]

The etiology of cap polyposis is unclear, with evidence for infectious and inflammatory causes involving mucosal ischemia and T-cell-mediated inflammation as well as mechanical causes, such as abnormal bowel motility and trauma to the rectal mucosa from straining. A common first step for the treatment of cap polyposis is instructing the patient to avoid straining during defecation. Treatment regimens similar to those for inflammatory bowel disease, such as topical or systemic steroids, infliximab, or aminosalicylates, have also been used.[5] Antibiotics to treat *Helicobacter pylori* have also been used. We present an extremely rare case of pediatric cap polyposis that was managed with wide-field endoscopic mucosal resection (W-EMR).

CASE REPORT

A 16-year-old adolescent boy had been having mucoid diarrhea, occasional rectal bleeding, and intermittent fecal incontinence for 6 years. A previous colonoscopy showed pseudopolyps in the rectum. Biopsies of these polyps revealed inflammation with granulation tissue. The initial laboratory evaluation revealed mild iron deficiency, anemia, and hypoalbuminemia.

Several of the larger polyps were removed with endoscopic polypectomy, and the patient's symptoms were managed with topical steroids and diphenoxylate-atropine. However, there was minimal improvement, and he was referred for surgical management. The surgeon then referred the patient to a pediatric gastroenterologist. A full workup for possible inflammatory bowel disease was performed with upper endoscopy, colonoscopy, and magnetic resonance enterography. There was no evidence of inflammatory

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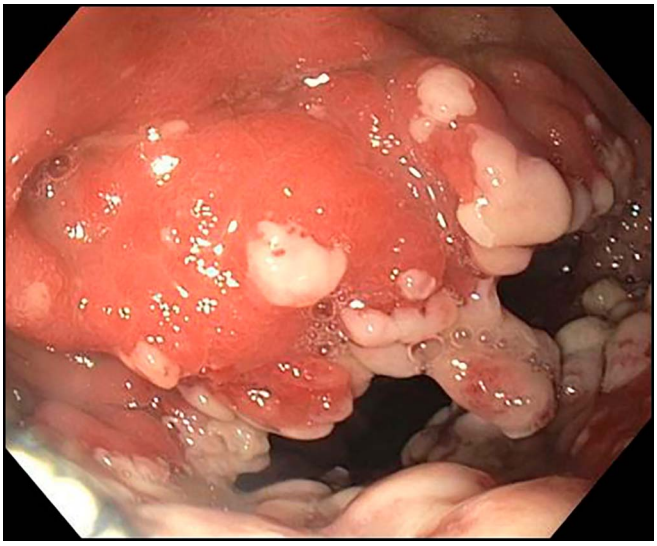

Figure 1. Initial colonoscopy showed multiple polyps with friable, white areas on the surface.

bowel disease, but there was marked polyposis in the rectum (Figure 1). The location and appearance of the lesions were suggestive of cap polyposis, and this was confirmed by biopsy (Figure 2). Tests for *H. pylori* were negative.

Neither the symptoms nor the polyposis resolved with antibiotic treatment (metronidazole only) and instruction to reduce straining during bowel movements. A repeated sigmoidoscopy showed diffuse 1–4 cm multilobulated polypoid lesions in the rectum (Figure 3). Endoscopic submucosal dissection (ESD) was initially considered but deferred because of the circumferential extent of the lesions and inadequate lifting after methylene blue/saline injection. Submucosal fibrosis was suspected given the lack of lifting; however, the sample for histology was not deep enough to demonstrate this. Thus, we performed polypectomy by W-EMR with multiple band ligations. For this procedure, we used a regular gastroscope with a distal cap attachment, which was removed when the banding device was placed. There were approximately 40 band resections, with near-complete removal of all the lesions (Figure 4). The lesions were focal and easily seen through the banding cap. The procedure took a total of 90 minutes, and there was no postprocedure bleeding, pain, or other adverse effects. The resected tissue specimen had signs of high-grade dysplasia away from the resection margins; however, we were unable to determine which polyp exhibited this feature.

On clinical follow-up, the patient no longer experienced mucoid diarrhea, lower gastrointestinal bleeding, or incontinence. A second-look procedure was performed 3.5 months later, which showed an almost entirely normal rectal mucosa with only a small area of nodularity (Figure 5), which was resected. This sample showed mild hyperplastic changes and crypt distortion, but no histologic features of cap polyposis. The patient was doing well without any symptoms on clinical follow-up 3 years after resection, but declined a follow-up sigmoidoscopy.

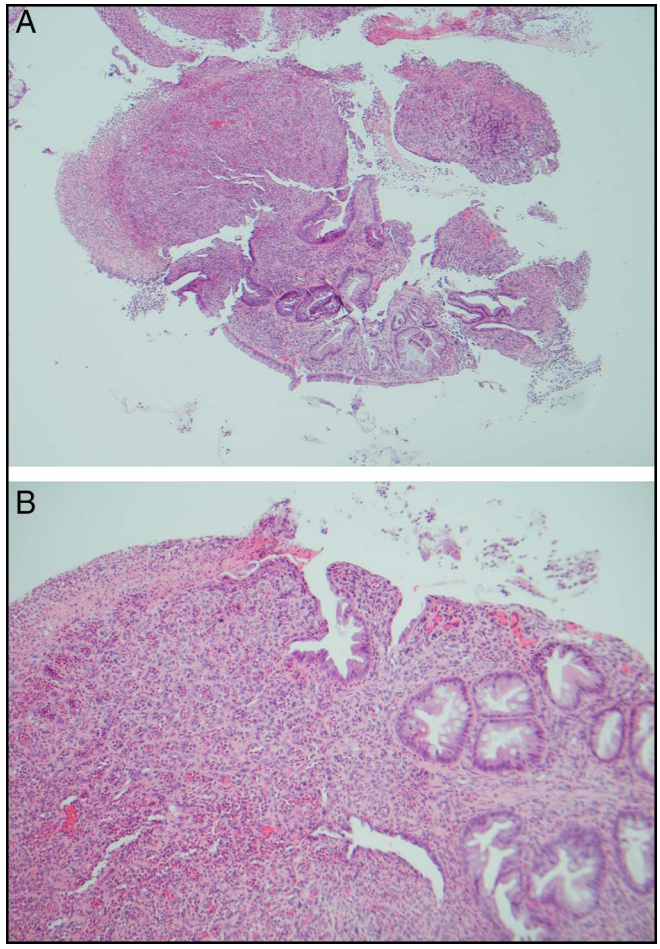

Figure 2. (A) Low-power magnification (2×; hematoxylin and eosin stain) showing hyperplastic and tortuous crypts with adjacent inflamed lamina propria. (B) Higher power magnification (10×; hematoxylin and eosin stain) demonstrating ulcerated granulation tissue and focal fibrinous exudates at the surface.

DISCUSSION

We present a rare case of cap polyposis that eluded diagnosis for several years and for which the primary symptoms were problematic diarrhea. Polypectomy with a band ligation-assisted W-EMR technique successfully alleviated the patient's symptoms.

Cap polyposis has histological similarities (fibromuscular obliteration and hyperplastic glands) to mucosal prolapse syndrome, which suggests it may be associated with excessive straining during defecation and an internal prolapse of the mucosal folds.[6,7] Indeed, the rectum is predominantly involved, and avoidance of excessive straining improves or eliminates symptoms in most, but not all, patients.[6,8] However, endosonographic differences between cap polyposis and mucosal prolapse syndrome have been noted by Gehenot et al[9] and Konishi et al.[10] Some cases of cap polyposis seem to have an infectious cause, as metronidazole is curative in some cases (29%).[11] Eradication therapy has been successful for all reported cases of *H. pylori*-associated cap polyposis.[11] There are no

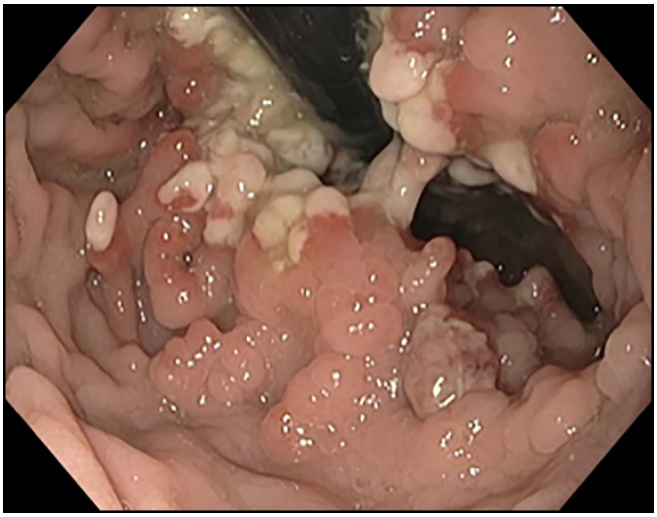

Figure 3. A repeated colonoscopy showed the extent of the polypoid lesions, which were nearly circumferential in the midrectum, with some of the polyps extending down close to the dentate line.

reports of *H. pylori*-associated cap polyposis in children. Li et al[6] described 1 child who had tested negative for *H. pylori* and, nevertheless, received triple therapy, but in whom symptoms continued to recur. The success rate for other treatments such as corticosteroids and aminosalicylates is low.[12,13] In our patient, topical steroid therapy had failed.

The outcomes for patients with cap polyposis vary from spontaneous remission[14] to recurrence after surgical resection.[3,9] There has been some reported success with endoscopic treatment, such as polypectomy and EMR.[5,15] However, surgical resection has often been pursued for unresponsive disease.[3,10,16] Because there have been no reports of malignant transformation in cap polyposis, surgical resection may be unnecessary. As a less

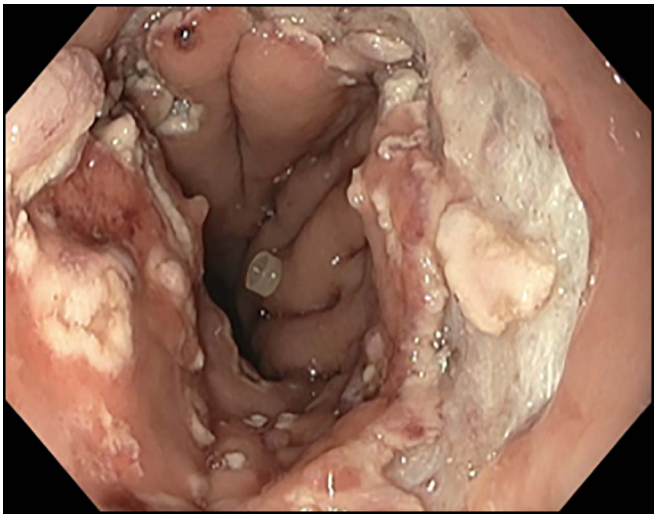

Figure 4. During W-EMR resection, an area of confluent mucosectomy was seen in the rectum. A repeated ligation EMR was performed at 40 separate sites. W-EMR, wide-field endoscopic mucosal resection.

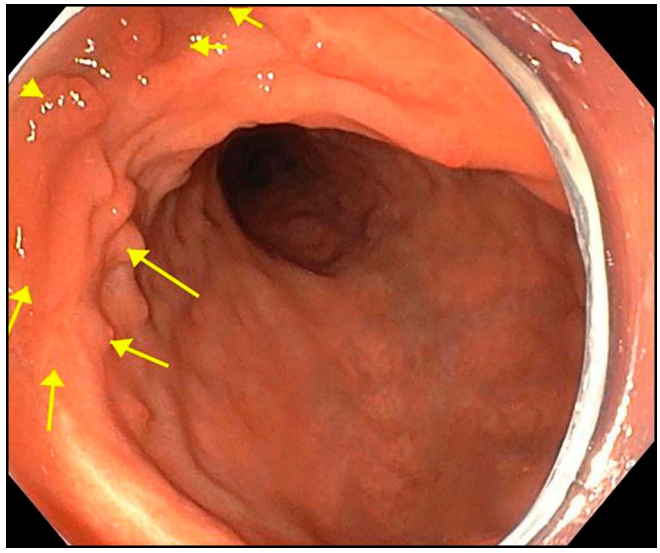

Figure 5. Follow-up colonoscopy showed nearly complete resolution of the polyps, with only a small residual area of nodularity (arrows).

invasive alternative, Murata et al[17] suggested endoscopic management with ESD en bloc excision. In the present case, the extent of submucosal fibrosis and the circumferential nature and localization of the lesions would have made ESD very challenging. Furthermore, we were able to preserve the normal intervening mucosa by performing multiple ligations, which can limit postresection stenosis that may be encountered with circumferential ESD.

Ligation EMR is a well-established technique to create a pseudopolyp using a band ligator, followed by electrocautery snare resection.[18,19] Although ligation EMR is most commonly used in the esophagus for nodular Barrett's esophagus, it can be safely performed for sessile polyps of the rectum.[20] However, its use in the colon should be limited to the rectum, and it should not be performed in the small intestine or large intestine above the peritoneal reflection. Ligation EMR enables precise targeting of lesions, particularly near the dentate line.[20] W-EMR is a reasonable alternative to ESD if en bloc resection and preservation of tissue margins are not required, and the procedure can be completed in a shorter time than ESD.[20]

In summary, cap polyposis may be successfully managed by W-EMR with successful resolution of symptoms. In adult patients, testing for *H. pylori* is recommended because, if present, eradication therapy seems effective. However, no pediatric *H. pylori*-associated cases have been described to date. For cases of cap polyposis in which medical management fails, W-EMR may be considered.

DISCLOSURES

Author contributions: S. Tomar preformed the literature search and prepared, edited, and reviewed the manuscript. M. Maksimak revised the article for intellectual content. D. Diehl

provided the images, critically reviewed the article, and is the article guarantor. All authors approved the final version of the manuscript.

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Informed consent was obtained for this case report.

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