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CASE REPORT

Cap polyposis in children: case report and literature review

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Abstract

Purpose Cap polyposis is a benign disease of the colon, characterized by inflammatory polyps with a "cap" of inflammatory granulation tissue. Its prevalence is very low, especially in children.

Methods and results A 16-year-old girl presented with frequent bowel movements with mucous discharge and bloody stool, leading to the initial suspicion of chronic inflammatory bowel disease. Results of further investigation by endoscopy and histological examination were consistent with a diagnosis of cap polyposis. Treatment with systemic steroids resulted in symptom improvement.

Conclusion A review of the literature shows that cap polyposis can occur at any age, including childhood, with a slight predilection for the male gender. Rectal bleeding and rectal polyps are consistent features in all reported cases. Other typical symptoms include constipation, diarrhea, and abdominal pain. Symptoms may be very similar to those of chronic inflammatory bowel disease, the most important differential diagnosis. The cause of cap polyposis is still unclear, and specific therapy has not as yet been established. Conservative therapeutic measures should be preferred, especially in children.

Keywords Cap polyposis · Rectal polyp · Inflammatory polyp · Rectal prolapse · Children

Introduction

Cap polyposis is a rare benign disease of the colon, characterized by inflammatory polyps with a "cap" of inflammatory granulation tissue. The lower rectum is most frequently affected. First described in 1985 by Williams et al. in 15 patients [1], cap polyposis has been reported in fewer than 100 cases at present.

Its origin remains unclear, although various causes are hypothesized—mechanical to infectious to inflammatory. Characteristic clinical symptoms are constipation, mucous discharge, diarrhea, abdominal pain, and rectal bleeding [2]. Inflammatory bowel disease is a frequent differential diagnosis owing to similarities in clinical and endoscopic presentation. Various therapeutic regimens have been described, but no specific therapy can be considered established.

The disease usually affects adults at a median age of 50 and is rarely described in children. We present the case of a child diagnosed with cap polyposis and review the literature, summarizing and analyzing the available data in children.

Case report

A 16-year-old girl presented with frequent bowel movements with mucous discharge and bloody stool. Patient history revealed the presence of symptoms over 3 years. The patient suffered from frequent stool emptying (approximately every 2 h), blood and mucous deposits on the stool, episodic constipation, and abdominal pain. There was no weight loss.

At presentation, several examinations had already been performed: Colonoscopy 2.5 years previously was without pathological finding. Increased calprotectin value had led to the suspicion of chronic inflammatory bowel disease. Sorbitol intolerance was found, but dietary correction did not improve symptoms. Therapy with local mesalazine application to treat proctitis was likewise ineffective. A recent flexible rectoscopy had found a circular polypoid mass extending from the anal

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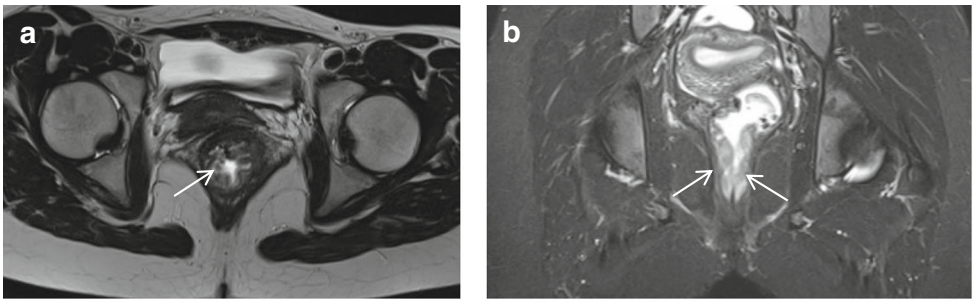

Fig. 1 Axial (**a**) and coronal (**b**) MRI demonstrating the polyp (white arrow)

canal to 10 cm from the anal verge. Histological examination of biopsies at an external hospital revealed the presence of exuberant granulation tissue closely mimicking a pyogenic granuloma.

At admission, digital rectal examination confirmed the presence of a circular polypoid mass in the rectum extending into the upper anal canal. Flexible colonoscopy revealed its extension to 8 cm from the anal verge. Results of laboratory tests (blood count, CRP, electrolytes, albumin) were unremarkable.

The patient underwent MRI of the pelvis (Fig. 1). The lesion presented as an unclear mass in the distal rectum reaching into the anal canal with short missing fat lamella between the vagina and the tumor. Repeat colonoscopy with biopsies was initiated (Fig. 2). Histopathologically, no conclusive diagnosis could be made owing to limited biopsy size raising uncertainty regarding the whole mass. Examination under anesthesia with excision of more tissue was carried out.

Histopathological examination showed extensive surface erosion of the rectal mucosa covered by a prominent exudative cap composed of fibrinous exudate admixed with inflammatory cells and mucous secretion (Fig. 3). The crypts showed prominent cystic dilation with variable goblet cell hyperplasia and a serrated appearance. Both intracryptic and interstitial mucin was seen. The lamina propria was replaced by capillary-rich florid granulation tissue characteristic of cap polyposis, and in other areas fibromuscular obliteration of the lamina propria was seen. The histological features were essentially very reminiscent of those in rectal mucosal prolapse (Fig. 3). Thus, cap polyposis was diagnosed.

The patient was first investigated for *Helicobacter pylori* infection, which was not confirmed [3]. Results of the urea breath test as well as immunohistochemical examination of cap polyposis rectal tissue were negative for *Helicobacter pylori*. Therefore, no eradication therapy was performed, but systemic steroid therapy was initiated (50 mg/day, reduced by 10 mg/day each succeeding week until a dosage of 20 mg/day was reached, after which the dose reduction was 5 mg/day each week). This markedly improved the patient's symptoms, but endoscopic findings remained (Fig. 4). On endosonography, the known polypoid mass showed no evidence of wall-transcending growth (Fig. 4b).

Discussion

The rarity of cap polyposis, especially in children, prompted us to report our 16-year-old patient and undertake a literature review. A PubMed search of the key phrase "cap polyposis," covering the time from 1993 to 2017, found 41 reports of 74 cases. Of these, 15 (20%) affected children. These cases were reviewed and patient data analyzed regarding age, gender, clinical and endoscopic presentation, differential diagnosis, treatment and outcome (Table 1) [4–8].

Table 2 summarizes the characteristics of cap polyposis in children. Based on the reviewed cases, it appears to affect

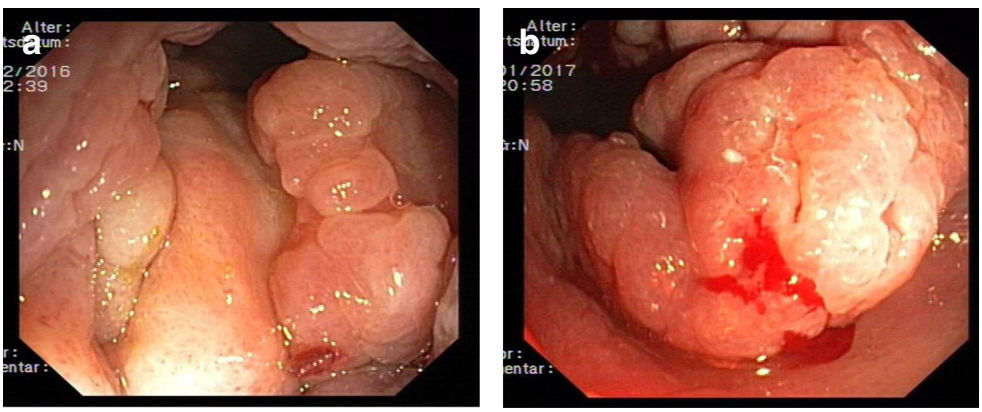

Fig. 2 Colonoscopic images with cap polyposis before (**a**) and after biopsy (**b**)

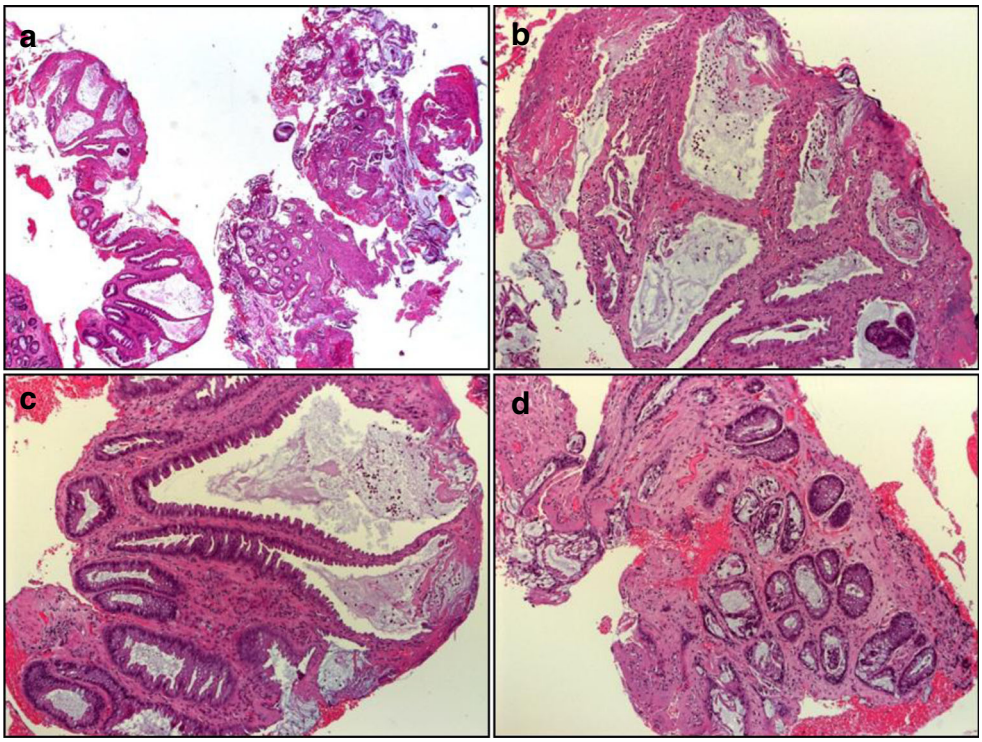

Fig. 3 Histopathological findings. **a** Prominent polypoid lesions seen at low power with extensive surface erosion and crypt dilatation. **b** Crypt dilatation and prominent mucous oversecretion within the crypts. **c** Prominent cystic dilation with variable goblet cell hyperplasia and serrated appearance. Note prominent exudative cap composed of fibrinous exudate admixed with inflammatory cells and mucous secretion. **d** Fibromuscular obliteration of the lamina propria by capillary-rich granulation tissue mimicking rectal prolapse findings

patients of any childhood age (range 1–17 years, median 12 years). The youngest affected patient was 11 months [8]. There is a male predominance, but without statistical significance (10/16, 63%). In contrast, among adults, the female gender (73%) predominates [9].

All children with cap polyposis presented with a common symptom—rectal bleeding (100%). Constipation (53%), diarrhea (40%,), and abdominal pain (40%) were less frequent. In comparison, most common symptoms among adults are mucous discharge/diarrhea (87%), bloody stool (33%), weight loss (10%), abdominal pain (10%), and tenesmus (10%) [9]. Protein-losing enteropathy represents another, albeit less common, symptom.

Cap polyposis is characterized by polyps with a "cap" of fibrinopurulent exudate. Polyps can vary in size (from a few millimeters to several centimeters), morphology (polypoid, ulcerative, or flat), number, and location. Endoscopic features in children show that cap polyposis is almost always confined to the rectum, with a rare extension to the anal canal or the sigmoid colon (14% each). Findings in the colon [10] or even in the stomach [11], as described in adults, have not been reported in children. In most cases, multiple manifestations are present (75%).

Histologically, cap polyps show typical and reproducible characteristics, as were also present in our patient. These are closely similar to and mimic findings in rectal mucosal prolapse, which led several authors to consider cap polyposis in that spectrum [6, 8, 12].

Inflammatory bowel disease is also an important differential diagnosis in children because of the similarity in

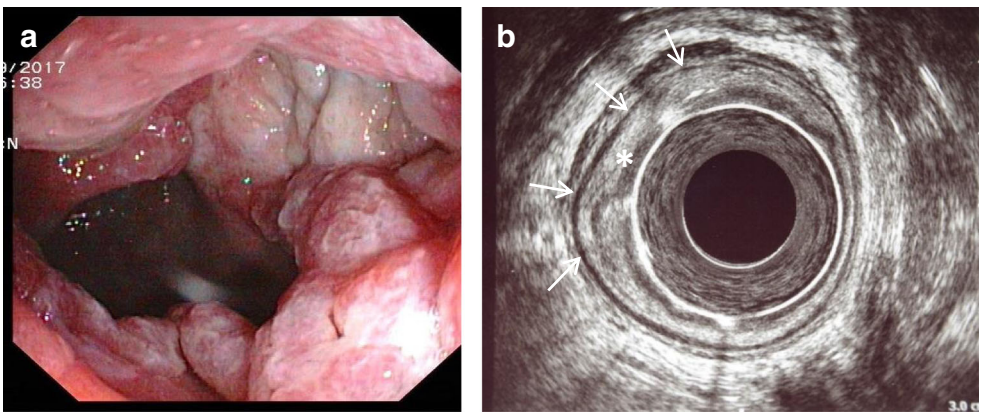

Fig. 4 Colonoscopic (**a**) and endosonographic (**b**) image with cap polyposis after therapy with systemic steroids. Polypoid mucosal mass is marked by asterisk, submucosal layer (white arrows) is not perforated

Table 1 Reported cases of cap polyposis in children

Case	Ref.	Year	Age	Gender	Clinical presentation	Localization /solitary vs. multiple lesions	Initial diagnosis	*Helicobacter pylori*	Treatment	Outcome
1	Shimizu et al.	2002	12 years	Female	- Mucous, bloody diarrhea	Rectum/multiple	Ulcerative colitis	Not reported	Metronidazole	Resolved
2	Ng et al.	2004	15 years	Male	Not reported	Not reported /multiple	Not reported	Not reported	Instruction to avoid straining + high-fiber diet Anterior resection	Failed Resolved
3	Li et al.	2013	5 years	Male	- Rectal bleeding	Rectum/solitary	Not reported	Not reported	Polypectomy + stool softener	Resolved
4	Li et al.	2013	13 years	Male	- Constipation - Rectal bleeding	Rectum/multiple	Not reported	Not reported	Polypectomy + stool softener	Resolved
5	Li et al.	2013	8 years	Male	- Constipation - Rectal bleeding	Rectum/solitary	Not reported	Not reported	Polypectomy + stool softener	Resolved
6	Li et al.	2013	8 years	Male	- Constipation - Rectal bleeding	Rectum/solitary	Not reported	Not reported	Polypectomy + stool softener + metronidazole	Resolved
7	Li et al.	2013	15 years	Female	- Diarrhea - Rectal bleeding	Rectum/multiple	Not reported	Not reported	Sulfasalazine	Resolved
8	Li et al.	2013	15 years	Male	- Diarrhea - Constipation - Abdominal pain - Rectal bleeding	Rectum/multiple	Not reported	Not reported	Polypectomy + stool softener + metronidazole	Recurrence
9	Li et al.	2013	10 years	Male	- Diarrhea - Abdominal pain - Rectal bleeding	Rectum/multiple	Not reported	Not reported	Polypectomy + stool softener + metronidazole	Recurrence
10	Li et al.	2013	11 years	Female	- Constipation - Abdominal pain - Rectal bleeding	Rectum/multiple	Not reported	Not reported	Polypectomy + stool softener + metronidazole	Recurrence
11	Li et al.	2013	13 years	Female	- Constipation - Abdominal pain - Rectal bleeding	Rectum/multiple	Not reported	Not reported	Polypectomy + stool softener	Recurrence
12	Li et al.	2013	14 years	Female	- Constipation - Abdominal pain - Rectal bleeding	Rectum + sigma/multiple	Not reported	Not reported	Polypectomy + stool softener	Recurrence
13	Li et al.	2013	17 years	Male	- Diarrhea - Rectal bleeding	Rectum + sigma/multiple	Not reported	Not reported	Polypectomy + stool softener	Lost to follow-up
14	Batra et al.	2016	16 years	Male	- Rectal bleeding	Not reported/multiple	Ulcerative colitis	Not reported	Mesalamine Steroids Polypectomy	Failed Failed Resolved
15	Kim et al.	2017	11 months	Male	- Rectal prolapse - Bloody stool - Epidermal nevus in the sacral area	Rectum + anus/multiple	Cap polyposis	Negative	Transanal polypectomy Recurrence: Mesalazine enema	Recurrence Resolved
16	Present case	2017	16 years	Female	- Mucous bloody diarrhea Sometimes: - Constipation - Abdominal pain	Rectum + anus/solitary	Inflammatory bowel disease (external diagnosis)	Negative	Mesalazine Steroids	Failed Improved

Table 2 Characteristics of cap polyposis in children*

Age (in years) mean [range]		12 [1–17]
Gender	Female	6/16 (38%)
	Male	10/16 (63%)
Clinical presentation	Diarrhea	6/15 (40%)
	Rectal bleeding	15/15 (100%)
	Constipation	8/15 (53%)
	Abdominal pain	6/15 (40%)
	Rectal prolapse	1/15 (7%)
	Epidermal nevus	1/15 (7%)
Localization	Rectum	14/14 (100%)
	Anus	2/14 (14%)
	Sigma	2/14 (14%)
Lesions	Solitary	4/16 (25%)
	Multiple	12/16 (75%)
Initial diagnosis	Inflammatory bowel disease	3/4 (75%)
	Cap polyposis	1/4 (25%)
Helicobacter pylori	Negative	1/1 (100%)
Treatment/outcome	Instruction to avoid straining + high-fiber diet	0/1 (0%)
	Anterior resection	1/1 (100%)
	Metronidazole	2/5 (40%)
	Polypectomy	5/11 (45%)
	Mesalazine	1/3 (33%)
	Sulfasalazine	1/1 (100%)

*Based on literature review of 15 reported cases from 1993 to 2017 and the present patient

clinical and endoscopic presentation. Indeed, in 75% of the reports in children, this was the initial diagnosis. Severe pseudomembranous colitis/proctitis is another consideration as it may mimic cap polyposis, underlining the importance of a carefully obtained clinical history and endoscopic evaluation [13].

Optimal targeted therapy is not as yet established because the cause of cap polyposis remains unclear. There are several hypotheses. The histological similarities (fibromuscular obliteration, hyperplastic glands) with mucosal prolapse syndrome have suggested disturbed intestinal motility, assuming excessive straining and an internal prolapse of the mucosal folds with formation of polypoid changes [6, 8, 12]. Further supporting this hypothetical cause are a predominant involvement of the rectum and the report of cases in which prevention of excessive straining during defecation has led to a cure or symptom improvement [6, 14]. Our patient also reported episodes of constipation. However, there are patients with cap polyposis with no history of straining and constipation, and the rate of healing through strain-avoidance is quite low. Furthermore, differences in endosonographic patterns between cap polyposis and mucosal prolapse syndrome were found by Gehenot et al. [15] and Konishi et al. [10].

Another suggested cause is an infectious component. In some cases, treatment with metronidazole led to a cure, although at a low rate (29%) [3]. Also, an association of cap polyposis with the presence of *Helicobacter pylori* infection has been described, and in these cases eradication therapy has been successful in up to 100% [3].

Further therapeutic approaches are steroids, TNF-alpha, and aminosalicylates, assuming that an inflammatory component could be the cause [16]. Reported success rates are low [5, 17]. In our patient, local mesalazine therapy had failed.

The natural course of cap polyposis is largely unknown. Reports range from spontaneous remission [18] to recurrence after surgical resection [2, 15]. Owing to the unknown pathogenesis, the varying course of the disease, and different treatment options with uncertain outcome, therapy remains challenging, as seen in our analysis of therapeutic and outcome data in children (Tab. 2). As yet there are no data in children regarding *Helicobacter pylori* eradication, which has been encouraging in adults, Li et al. described one child who had tested negative for *Helicobacter pylori* and nevertheless received triple therapy, but in whom symptoms continued to recur [6].

In summary, in adults, polypectomy should be performed to alleviate symptoms when possible. For medical management, testing for *Helicobacter pylori* is recommended because very good results are described for eradication therapy. A treatment trial with steroids seems to be useful. In children, however, conservative measures are preferred. If the disease persists or recurs despite medical treatment, surgical resection can be considered [5].

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

Informed consent We obtained written informed consent for the patient reported herein.

Statement of human rights For this type of study, formal consent is not required.

Statement on the welfare of animals This article does not contain any studies with animals performed by any of the authors.

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