Ulcerative colitis with inflammatory polyposis in a teenage boy: A case report

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Abstract

Ulcerative colitis in addition to inflammatory polyposis is common. The benign sequel of ulcerative colitis can sometimes mimic colorectal carcinoma. This report describes a rare case of inflammatory polyposis with hundreds of inflammatory polyps in ulcerative colitis which was not easy to distinguish from other polyposis syndromes. A 16-year-old Chinese male suffering from ulcerative colitis for 6 mo underwent colonoscopy, and hundreds of polyps were observed in the sigmoid, causing colonic stenosis. The polyps were restricted to the sigmoid. Although rectal inflammation was detected, no polyps were found in the rectum. A diagnosis of inflammatory polyposis and ulcerative colitis was made. The patient underwent total colectomy and ileal pouch anal anastomosis. The patient recovered well and was discharged on postoperative day 8. Endoscopic surveillance after surgery is crucial as ulcerative colitis with polyposis is a risk factor for colorectal cancer. Recognition of polyposis requires clinical, endoscopic and histopathologic correlation, and helps with chemoprophylaxis of colorectal cancer, as the drugs used postoperatively for colorectal cancer, ulcerative colitis and polyposis are different.

Key words: Ulcerative colitis; Inflammatory polyposis; Teenager; Colorectal cancer

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Core tip: This case report describes ulcerative colitis with inflammatory polyps in a teenage boy. The macropathology of inflammatory polyps excised from the colon was similar to that of familial adenomatous polyps and hyperplastic polyps. In this article, we discuss the difficulties in distinguishing inflammatory polyposis from similar polyps and emphasize the importance of the chemoprophylaxis of colorectal cancer developed from ulcerative colitis and polyposis.
INTRODUCTION

Ulcerative colitis (UC) is one of two major types of inflammatory bowel disease (IBD); the other is Crohn’s disease (CD). The age of onset follows a bimodal pattern, with a major peak at 15-25 years and a smaller peak at 55-65 years, although the disease can occur at any age [1]. Inflammatory polyps are usually found in the setting of severe inflammatory diseases such as IBD, and carcinoma can occur in inflammatory polyps, especially unusual inflammatory polyps in complex formations. Here we present the case of a 16-year-old male with inflammatory polyposis (IP) in addition to UC, and describe its appearance on colonoscopy and gross specimen following surgery. To the best of our knowledge, such a severe condition at such a young age is rare, and it is necessary to distinguish the polyposis in this case from other polyposis syndromes.

CASE REPORT

A 16-year-old Chinese male suffering from recurrent abdominal pain and diarrhea with mucosanguineous feces for six months was referred to our department on September 18, 2012. The patient had occasional fever with dark red stool and stench on one or two occasions. He showed no obvious weight loss, had no relevant family history, and did not smoke or drink alcohol. Six months previously he had been diagnosed with UC based on colonoscopy (Figure 1A) and histology of biopsy (Figure 1B, C). At that time, physical examination showed no obvious abdominal abnormality. Laboratory examinations showed C reactive protein (CRP) of 19.0 mg/L, hemoglobin (HGB) of 63 g/L, and platelets (PLT) of $503 \times 10^9$/L. The patient received mesalazine for six months, but the symptoms recurred.

Six months later on September 21, 2012, a second colonoscopy was performed, revealing a large number of polyps in the sigmoid lumen, mucosal swelling, friability, erosions, loss of vascular pattern, and substantial superficial punctate hemorrhage. The large number of polyps had also resulted in stenosis in the sigmoid lumen (Figure 2A). The lens was unable to pass through the stenosis into the enteric cavity. The rectal mucosa showed inflammation and ulcer formation, but no polyps.

The patient underwent a total colectomy and ileal pouch anal anastomosis (IPAA) on October 7, 2012. Macroscopic examination of the resected colon revealed a large number of diffuse inflammatory polyp-like protrusions (more than a few hundreds) at both ends of the excision. Polyp diameter ranged from 0.3 to 0.8 cm. Histological analysis showed inflammatory polyposis (Figure 2B-D), and no granulomatous, adenomatous or malignant changes were noted. After surgery, the patient’s condition improved and he was discharged on postoperative day 8. Examination was performed six months later, and an increase in stool frequency was observed. The boy’s quality of life was normal, and acute and chronic complications such as bleeding, pelvic abscess, pouchitis, pouch failure, intestinal obstruction, and chronic pelvic infection were not found.

DISCUSSION

Inflammatory polyps are often found in inflammatory diseases of the colonic mucosa, such as UC in remission, and they may produce symptoms of pain [2], and obstruction [3], especially giant polyps [4]. In this case, the patient developed UC at an early age which was quickly complicated by severe inflammatory polyposis, which is unusual in UC. As seen in the endoscope image, numerous polyps were mainly located in the sigmoid which led to stenosis, and there was no histological evidence of neoplasm or CD which may have resulted in stenosis of the lumen (Figure 2B-D). According to endoscopic and surgical findings, it is necessary to distinguish inflammatory polyposis from other polyposis syndromes.

Another type of polyposis is familial adenomatous polyposis (FAP) which is characterized by the presence of hundreds to thousands of adenomatous polyps throughout the colon. The World Health Organization (WHO) diagnostic criteria for FAP are as follows: (1) 100 or more colorectal adenomas or (2) a germline mutation of the APC gene; or (3) a family history of FAP and at least one of the following: epidermoid cysts, osteomas, and desmoid tumor. There appears to be no significant ethnic or racial differences in the incidence of FAP. In this case, the gross findings in the excised specimen were similar to FAP, which confused the diagnosis. The potential relationship between FAP and UC is not clear. A 50-year-old man with no known history or symptoms of IBD presenting with filiform polyposis involving the entire colon, clinically mimicking FAP, and showing histologic features similar to neuromuscular and vascular hamartoma of the small bowel was reported [5]. Leal and colleagues found that patients with UC had significantly higher protein levels of Bax, APAF-1, and Caspase-9 than patients with FAP [6]. The average age of onset of polyposis in FAP is 16 years, which was the age of our patient. However, the histologic appearance of the resected specimen did not match the criteria for FAP (Figure 2B-D).

Hyperplastic polyposis (HP) is another type of polyposis. HP is usually diagnosed in individuals in their 40’s to 60’s, although it has been reported in patients as young as 11 years [7]. The syndrome and its inherent risk of malignant disease should be considered when polyps are numerous (more than 20, i.e., polyposis), large ($> 1$ cm), and proximally located (especially if more than five are proximal to the sigmoid colon), and especially when there are serrated adenomas [8,9]. A family history of HP is uncommon, but colorectal cancer (CRC) in a first-degree relative of a patient with HP is common and reflects inherited increased risk, perhaps associated with the putative heterozygous state [10].

There are similar characteristics in UC, FAP and HP, respectively. Firstly, individuals with these diseases are
clearly at increased risk of CRC. FAP is an autosomal-dominant CRC syndrome that can be caused by a germline mutation in the adenomatous polyposis coli (APC) gene on chromosome 5q21[11]. For patients who inherit the FAP mutation, there is a virtually 100% risk of colon cancer[12,13]. Colon cancer will develop in some patients as early as pre-teenage years[14]. The proportion of FAP patients with CRC who are under 20 years old is 2%-15%[15]. In addition, FAP is associated with an increased risk for the development of other malignancies, such as desmoid tumors, lymphoma, adrenal cancer, gastric cancer and ileal adenomas. A high incidence of duodenal polyps has also been described in FAP patients (79.3%)[16].

Recent studies have proposed that, large right-sided sessile “serrated” hyperplastic polyps are prone to oncogenetic and epigenetic changes leading to genetic instability and neoplasia[17,18]. HP will progress to adenocarcinoma through a “serrated neoplastic pathway” and a B type Raf kinase (BRAF) proto-oncogene mutation[19]. About 30% of CRCs develop through this pathway[20]. However, BRAF mutations were not present.

Figure 1  Colonoscopy and histologic findings in the early onset of colon inflammation. A: Inflammation in the colon, no polyps were found in the colon; B: Mucosal epithelial necrosis, destruction, distortion and branching of lamina propria glands. The distance between the glands in some regions was increased and goblet cells decreased significantly; a large number of lymphocytes and plasma cells infiltrated the lamina propria, there were neutrophils in crypts, and lymphocytes increased significantly in the base of the mucosa; a large number of lymphocytes and plasma cells infiltrated the submucosa; C: Lamina propria glands were distorted and branched accompanied by crypt abscesses and parts of the glands showed atrophy. The distance between the glands in some regions was increased, in which goblet cells were decreased. Inflammation in the lamina propria was uniform, with infiltration of numerous lymphocytes and plasma cells, and stromal vessels were significantly dilated and congested (HE staining).

Figure 2  Colonoscopy and histologic finding 6 mo later. A: Diffuse congestion and edema in the mucosa accompanied by focal hemorrhage and exudation, showing the different shapes, sizes, erosive lesions and superficial ulcers, and most of the regional mucosal hyperplasia was granular with pseudopolyp formation; B-D: Formation of necrosis and shallow ulcers in part of the mucosal epithelial and glands in the lamina propria were distorted and branching. Several glands showed atrophy and goblet cells were decreased. Neutrophils infiltrated the lamina propria. The stromal blood vessels were significantly dilated and congested (HE staining).
in any of the polyps of patients with hyperplastic/serrated polyposis in addition to IBD[22]. These findings suggest the possibility of another pathway related to carcinogenesis in IBD. The genetic abnormalities in HP also include oncogenes and tumor suppressor genes, especially abnormalities in KRAS and TGFBR2, and loss of chromosome 1p.

UC is also associated with an increased risk of CRC, depending on age at diagnosis, especially in those less than 15 years of age, and is also dependent on the extent of disease at diagnosis[23]. The risk of CRC in patients with UC is approximately 7%–14% by 25 years of age, and the overall incidence rate of CRC is 1.67 per 1000 patient-year[24]. A recent study showed that the risk of developing CRC in patients with UC has steadily decreased over the last six decades, but the extent and duration of the disease has increased this risk[25]. The available data indicate a similar proportion of CRC in FAP and UC patients[29,30]. Whether patients with FAP in addition to UC have a higher risk of CRC is unknown and requires further investigation.

Secondly, abdominal pain and bloody diarrhea with or without mucus are common symptoms of UC and FAP. For patients with UC in addition to other polyposis, it is possible that the early symptoms of UC masked the symptoms of polyposis, however, in some cases, UC is an initial symptom of polyposis, and vice versa. It was reported that late-onset UC can present as filiform polyposis, although there were no associated diverticula, inflammatory lesions or adenomas on endoscopy and the histology of intervening mucosa was strongly suggestive of UC in remission[29]. Thus, there may be a relationship between FAP and UC which needs to be clarified. Another report suggested a link between IBD and FAP as the offspring of an FAP patient suffered from IBD[26]. In this case, however, none of his members suffered from IBD or other polyposis syndromes, and his polyps were diagnosed after inflammation was observed.

There are some differences among the three types of lesions mentioned above. UC, HP and FAP are all closely related to CRC, however, the chemoprophylaxis for CRC is important, but is contraindicated in UC and neoplastic polyposis. For CRC and some neoplastic polyposis, non-steroidal anti-inflammatory drugs (NSAIDs) are beneficial, but are harmful in UC. NSAIDs play a role in postponing surgery in patients with mild colonic polyposis, in patients with rectal polyposis after surgery, and as an adjunct to endoscopic surveillance; the use of any NSAID regardless of type is associated with a reduced risk of adenomatous polyps[27]. However, the risk is increased in CD and UC[28] and NSAIDs have been reported to induce irreversible exacerbation of IBD[29,30]. As a chemopreventive agent in CRC, the effectiveness of aspirin in FAP is unclear[24]. Researchers have found that the use of aspirin and COX-2 inhibitors was not associated with HP risk[31]. In UC patients, the use of 5-aminosalicylic acid (5-ASA) can reduce the risk of CRC[32], however, 5-ASA appears to have no effect on reducing the number or shrinking the size of polyps in the APCmin mouse model of FAP[33].

In conclusion, a teenage boy with UC in addition to polyposis with a possible higher risk of CRC is described. Thus, the recognition of polyps requires the correlation of clinical, endoscopic and histopathologic examinations. It is important to make an accurate diagnosis as the chemoprophylaxis for CRC is contraindicated in patients with neoplastic polypos or pseudopolyps. Endoscopic surveillance is necessary for patients who have undergone colectomy.

**COMMENTS**

**Case characteristics**

Hundreds of polyps were found at the onset of ulcerative colitis in a teenage boy.

**Clinical diagnosis**

Ulcerative colitis complicated by inflammatory polyps: Large numbers of polyps resulted in stenosis in the sigmoid lumen.

**Differential diagnosis**

Endoscopic and surgical examinations were required to distinguish inflammatory polyposis from familial adenomatous polyposis and hyperplastic polyposis.

**Laboratory diagnosis**

Laboratory diagnosis was ulcerative colitis with inflammatory polyps.

**Imaging diagnosis**

Colonoscopy images showed hundreds of polyps in the sigmoid causing colonic stenosis.

**Pathological diagnosis**

Pathological diagnosis indicated inflammatory changes in the colon.

**Treatment**

The patient underwent a total colectomy and ileal pouch anal anastomosis.

**Term explanation**

There are no uncommon terms in this case report.

**Experiences and lessons**

Some inflammatory polyposis syndromes are similar to familial adenomatous polyposis and hyperplastic polyposis. These disorders should be carefully distinguished and accurately diagnosed to ensure an appropriate therapeutic regimen.

**Peer review**

Inflammatory polyposis is not novel in ulcerative colitis but the review of the literature is sound and complete for this case report.

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