A case of the serious Crohn’s disease in pediatric patient

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Abstract

Crohn’s disease (CD) and ulcerative colitis (UC) represent the two major forms of inflammatory bowel disease (IBD), which is a chronic, immune-mediated disease of the gastrointestinal tract that develops in genetically susceptible individuals [6]. CD primarily affects the bowel, but can also involve the musculoskeletal system, skin, and eyes [10]. CD manifests during childhood or adolescence is up to 25% of patient, but recent observations indicates the decrease of the age of diagnosis and the increase of aggravation of the disease [5]. Because the symptoms of CD are similar to other intestinal disorders, such as irritable bowel syndrome and UC, it can be difficult to diagnose. The most common symptoms of CD are abdominal pain, often in the lower right area, and diarrhea. Rectal bleeding, weight loss, arthritis, skin problems, and fever may also occur. Varied phenotypic features and a spectrum of clinical severity are observed in children with the disease, as among adults, but the potential for linear growth impairment is unique to pediatric patients [4]. The gold standard of IBD diagnosis is histological evaluation [2, 12]. The most common complication is blockage of the intestine, which occurs because the disease tends to thicken the intestinal wall with swelling and scar tissue, narrowing the passage. CD may also cause sores, or ulcers, that tunnel through the affected area into surrounding tissues, such as the bladder, vagina, or skin. The areas around the anus and rectum are often involved. The fistulas are a common complication and often become infected. There is no known cure for CD. Treatment may include drugs, nutrition supplements, surgery, or a combination of these. The choice of the treatment depends on the location and severity of disease, complications, and the person’s response to previous medical treatments when treated for recurring symptoms [1, 14]. Two-thirds to three-quarters of CD patients will require surgery at some point in their lives. Invasive procedures become necessary when medications can no longer control symptoms. Surgery is used either to relieve symptoms that do not respond to medical therapy or to correct complications such as blockage, perforation, abscess, or bleeding in the intestine [8, 10].
In this case report we present the severe, refractory Crohn’s disease with complications and extraintestinal manifestations in 17 year old boy after total colectomy.

Case report

A 17-year old boy with severe, refractory Crohn’s disease diagnosed in 2005 has been under medical care of the Children’s Memorial Heath Institute since 2007. He had been already treated with all possible pharmacologic treatments, such as corticosteroids, immunomodulators (both azathioprine and methotrexat), anti-inflammatory salicylates (5-ASA) and antibiotics. The boy was hospitalized many times due to exacerbations of a disease. He was admitted to our Institute because of the lack of remission and refractoriness of his CD. The successive aggravations of a disease in our patient qualified him for biologic therapy. The boy was applied three doses of Infliximab in 2008, but he finally lost his response to the treatment. There were both clinical and endoscopic symptoms of deterioratin and exacerbation of a disease noticed, therefore the drug was withdrawn after all. Because the state of the boy was serious, we decided to administer Adalimumab, but after short period of improvement, the aggravation appeared again in our patient. He was referred to hospital and applied an increased dose of a biologic drug but with no clinical effect. Therefore, we resigned from the treatment due to lack of remission. During further hospitalizations the large lesions in the distal part of the colon, extending from the rectum to the left flexure of the colon were detected. The bowel was narrow, stiff and shortened (Fig. 1). The boy was finally qualified for surgery. The ileostomy was exteriorized (35 cm ahead of the ileocolic valve) in 2008. During hospitalization, a long term antibioticotherapy was applied together with the parenteral nutrition but no clinical effects were observed. The control endoscopic examinations, both gastro and colonoscopy, were performed, presenting gastritis and very severe Crohn’s disease. The patient was in poor general condition. The cachexy, fever and extensive perianal lesions remained despite the broad spectrum antibioticotherapy, immunosupresive treatment (both azathioprine and metotrexat) and total parenteral nutrition administration.

The decision of colectomy was made due to all therapeutic options depletion. The patient underwent total colectomy with removal of the anus and rectum together with exteriorization of the final ileostomy in July 2009. A large perianal fistula on the right side and 25 cm of the small intestine below ileostomy were removed (Fig. 1). There were no complications during the surgery. Parenteral nutrition was applied, replaced subsequently by the oral feeding. The nourishment of our patient was significantly improved. He was discharged from hospital in good general condition with prescription to continue his therapy in gastroenterology clinic.

Discussion

CD is a chronic inflammatory disorder that primarily affects the bowel (in contrary to UC-both small and large intestine as well as the esophagus), but can also involve the musculoskeletal system, skin, and eyes [10]. IBD most commonly begins during adolescence and early adulthood, but it also can begin during childhood and later in life. Our patient was diagnosed to have CD while he was 14 year old and presented typical symptoms for the disease, such as: the abdominal pain, bloody diarrhea, weight loss and the lack of appetite. The localization of the pathological inflammation was the large intestine and stomach (gastritis), there were no lesions within the small intestine. Growth impairment and associated pubertal delay are common complications of pediatric IBD, particularly CD [3]. The skeletal age of our patient was far below the norm for his age, he was also considerably malnurished. CD complications may be related or unrelated to the inflammation within the intestine (such as intestinal or extra-intestinal). Intestinal

Fig. 1 Colonoscopy pictures – a large perianal fistula on the left side and the narrow, stiff and shortened bowel on the right side
Complications of CD include obstruction and perforation of the small intestine, abscesses (collections of pus), fistulae, and intestinal bleeding. Extra-intestinal complications involve the skin, joints, spine, eyes, liver, and bile ducts [11]. In our case, there were both the stenosis within the rectum and the large perianal fistula. The symptoms and severity of CD vary among patients. Patients with mild or no symptoms as well as the ones in remission may not need treatment. However, there is no medication that can cure CD. Patients with the disease typically experience periods of relapse (worsening of inflammation) followed by periods of remission (reduced inflammation) lasting months to years. In our patient the periods of remission were very short in spite of intensive multidrug treatment, while during numerous relapses, which, in contrary, were very often, symptoms of disease worsen. The focus of treatment CD must be to reduce or eliminate symptoms, optimize nutritional status, promote normal growth and development, prevent complications and minimize the potential psychological effects of this chronic illness [9]. Treatment may include drugs, nutrition supplements, surgery, or a combination of these options. The pharmacotherapy includes: anti-inflammation drugs (5-ASA), cortisone or steroids, immune system suppressors (6-mercaptopurine, azathioprine), biologic therapy (Infliximab, Adalimumab), antibiotics and anti-diarrheal (diphenoxylate, loperamide, and codeine) together with fluid replacements. Nutritional supplements may be recommended, especially for children whose growth has been slowed. Special high-calorie liquid formulas are sometimes used for this purpose. A small number of patients may need to be fed intravenously for a brief time. Surgery becomes necessary when medications can no longer control symptoms. It is used either to relieve symptoms that do not respond to medical therapy or to correct complications such as blockage, perforation, abscess, or bleeding in the intestine [13]. In our case, the patient underwent all possible pharmacological treatments, including biologic therapy – both Infliximab and Adalimumab. However, none of the drugs seemed to be effective, as the patient presented steroid resistance and lost his response to both biologic agents. We also tried the nutritional therapy – the total parenteral nutrition was applied to the boy. But despite all these therapeutic possibilities there was no improvement of disease and the general state of our patient was poor. Therefore the decision of total colectomy was made. The boy underwent the surgery in 2009. There were no complications during procedure and the patient was discharged from hospital in good condition. Since the operation, the nourishment of our patient has significantly improved. There was even noticed the increase of his body mass. So far, the general state of our patient is well and he continues his therapy in gastroenterology clinic. In this case report, we presented the patient with severe, refractory Crohn’s disease, that finally led to total colectomy, which turned out to be the only treatment because no conservative therapies seemed to be effective.

References