Small Intestine Crohn’s Disease Presenting as Fever Mistaken for Adult Onset Still’s Disease

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Abstract

Crohn’s disease (CD) is not rare in recent years, but it is sometimes difficult to make a definite diagnosis particularly if it is in the small intestine. We report a patient with fever for 8 months whose disease was mistaken to be Adult onset Still’s disease. The patient was diagnosed small intestine Crohn’s disease at last by pathology. We want to emphasize that doctors should not forget small intestine Crohn’s disease when encountering an unidentified feverish patient, they should not diagnose a feverish patient of Adult onset Still’s disease at once. It is important to note that corticosteroids can conceal many diseases and they should not be considered lightly even if the patient is diagnosed with Adult onset Still’s disease.

Key words: Crohn’s disease, Adult onset Still’s disease

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Introduction

Crohn’s disease (CD) is a multifactorial polygenic disease characterized by chronic inflammation of the gastrointestinal tract (GIT) (1). It may involve any part of the GIT. The most frequent location of CD is in the terminal ileum and the colon. There are over 600,000 individuals with CD in North America, with up to 40,000 new cases being diagnosed each year, and double that amount at risk, based on a monozygotic concordance rate of 50% (2, 3). As such, an effective diagnosis can be made with the aid of ileocolonoscopy and biopsies in most cases.

Adult onset Still’s disease (AOSD) is the rare clinical condition characterized by high spiking fever, evanescent macular or maculopapular rash, poly- or oligoarthritis and leucocytosis (4). Numerous infectious agents have been proposed as potential inciting factors. Viruses such as Epstein-Barr virus, Coxsackievirus, adenovirus, influenza A, human herpes 6, hepatitis B, hepatitis C, and parvovirus B19 have been implicated in small case series (5). As we know, these two diseases are different, they have no relationship with each other. We will present a patient with fever for 8 months being diagnosed with small intestine Crohn’s disease. But during the course of treatment of this patient, some doctors mistook it for Adult onset Still’s disease. In this case, we want to tell, although Crohn’s disease is not rare, the diagnosis is sometimes difficult, some of its clinical findings are similar to Adult onset Still’s disease.

Case Report

A 47-year-old woman presented with 8 months of fever. Eight months earlier the patient suffered from fever, the fever happened in the afternoon and evening, and the highest temperature was 40.6°C, which is a temperature that could be normal after sweating. She had no headache, no cough, no chest pain, no abdominal pain, no nausea, vomiting or diarrhea. Then she went to local hospital to do test, CT of the lung and the head was normal. CBC, routine stool and urinary tests were normal, ANA was normal. Only the virus test showed cytomegalovirus infection. So she was diagnosed of infectious fever. After taking an antiviral agent, the patient had no fever for about a week. After that, the fever appeared again accompanied with ache of the knee joints and rash. So she went to another hospital to undergo a colonoscopy which showed point-like inflammation of the colon. Other tests such as CBC, routine stool and urinary tests, X-ray of the lung, electrocardiogram, ECT, and so on were all normal. At last she was diagnosed with Adult onset
Still’s disease. The doctor made a prescription of prednisone. During the prednisone administration, her temperature was normal most of the time, but sometimes her temperature went higher than 38°C. The rash and the ache of knee joints were relieved. Two months previously, after the patient stopped taking medicine, she had high fever again, the temperature could reach 40°C, so she came to our hospital.

The patient denied any history of hepatitis, TB, hypertension, diabetes, smoking, or drinking. After admission, her temperature was 39.6°C, pulse was 90 bpm, BP was 120/80 mmHg, respiration was 20 bpm. The auscultation of the lung and heart was normal. No lymph node enlargement, no tenderness of the abdomen, no rebound tenderness, no shifting dullness, no edema of the legs. Laboratory investigations demonstrated WBC 6.3×10^9/L, neutrophils percentage 57.50%, monocyte 15.6%, RBC 3.35×10^12/L, Hb 78.0 g/L, HCT 0.2680 ratio, PLT 30.5×10^9/L, stool occult blood (-), ESR 53 mm/h, CRP 27.3 mg/L, TBAB (-). Blood culture showed: cyst of the liver, cholecystitis, and polyp of the gall bladder. Bone marrow biopsy was normal. Radiography of the intestine showed the ileum limitations spasms change (Fig. 1). As the patient had a high fever for days, we considered that it was lymphoma and suggested her to undergo balloon enteroscopy, but the patient wanted an operation, according to the patient’s will, we did exploratory laparotomy. It showed the 75 cm long mucosa and the ileum 40 cm from the ileocecum was edematous and incrassated. The small intestine wall was thick, and longitudinal and transverse ulceration in an edematous mucosa induced a characteristic cobblestone appearance (Fig. 2). The diagnosis of exploratory laparotomy was tumor of the intestine. The pathology of the ileum showed small intestine Crohn’s disease (Fig. 3). The lesion of the intestine was resected and we asked her to take 5-ASA and to be followed up in clinic as an outpatient. We followed her up for 5 years, after the lesion of intestine was resected, she had no fever and no other symptoms again.

**Discussion**

Crohn’s disease is a chronic inflammatory process of unknown etiology affecting the gastrointestinal tract, uni- or multifocal, of variable severity, transmural (6), and it is not curable by clinical or surgical treatment. Associated or isolated extraintestinal manifestations can occur, affecting the skin, joints, eyes, liver, and urinary tract more commonly. It is characterized by recurring episodes of suppurative inflammation in any part of the gastrointestinal tract from the mouth to the anus. The inflammation is transmural and can result in strictures, microperforations, and fistulae. The in-
flammmation is non-contiguous and can thus produce skip lesions throughout the bowel. The incidence of CD in the US is 4/100000. As the disease is chronic, the prevalence is much higher: around 80-150/100000 (7, 8). The disease affects individuals of any age, but the diagnosis is more common in the second or third decades (9). CD can be categorized into three general types according to the predominant gross manifestation of the disease: stricturing disease, perforating disease, and inflammatory disease (10). CD of the upper gastrointestinal tract usually presents with nausea, vomiting, dysphagia, or odynophagia (11). The predominant symptom of small bowel CD is abdominal pain, which occurs in 90% of patients (12). Crohn’s involvement of the colon typically presents with diarrhea that may or may not be bloody.

In this case, although the patient was diagnosed of small intestine Crohn’s disease, she had no abdominal pain at all. We cannot be completely sure of the disease onset. So Crohn’s disease is always a chronic disease. Only when the clinic symptoms appear, the patient may come to the hospital, that is why it is always in advanced period when the patient comes for treatment. In this case, the patient only had fever at first, without any other symptoms, as the bone marrow biopsy and the blood culture were negative, we could first rule out common infectious disease and then in addition to fever, the patient had a rash and pain of both knee joints, these symptoms rendered the doctors confused with Adult onset Still’s disease. The mistake of the doctor was that Adult onset Still’s disease should first rule out other diseases. Throughout the follow-up period after resection, the patient had no fever and no rash or pain of joints again, that was why we could rule out AOSD. If the patient was diagnosed with AOSD, the fever would not disappear after resection. Thus the clinical manifestation of Crohn’s disease was not typical; this patient was presented with fever. So if we meet an unidentified febrile patient, we should think about Crohn’s disease. Because small intestine Crohn’s disease cannot be diagnosed by coloscopy, the radiographic abnormalities are often distinctive. So in this case, if we had not done radiography of the intestine, we still could not have found the lesion. As nowadays the diagnosis of small intestine Crohn’s disease can only depend on the pathology, but for a patient with fever, the doctor will not do routine radiography of the intestine, not to speak of abdominal laparotomy. Therefore, it is still a difficult question for a doctor to differentiate small intestine Crohn’s disease with only fever from other diseases with fever.

Here we had another question regarding the treatment. Corticosteroids and aminosalicylates are the common treatment for Crohn’s disease. Corticosteroids are only used to induce clinical remission. Aminosalicylates come in variety of preparations, each designed to deliver the drug to a particular intestinal segment (13). These include sulfasalazine and 5-aminosalicylic acid (5-ASA) derivatives. In this case, the doctor mistook the patient for Adult onset Still’s disease, and corticosteroids are the only treatment for this disease. But corticosteroids can also induce the clinical remission of Crohn’s disease. So when the patient stopped the medicine, the fever returned again. And the use of corticosteroids concealed the real disease. Therefore, this case taught us that doctors should not diagnose the patient with fever of Adult onset Still’s disease at once, even the patient had rash and pain of joints. These symptoms could also be seen in Crohn’s disease. It can be only diagnosed when all of other diseases can be ruled out. As corticosteroids would conceal a lot of condition of illness, doctors should not use it easily.
Conclusion

Crohn’s disease is not rare nowadays, but it is sometimes difficult to make a definite diagnosis especially when it is in the small intestine. The clinical appearance is not typical, sometimes only presents with fever. As coloscopy can not discover small intestine Crohn’s disease, also other routine examinations can not discover it, and sometimes it may involve the joints and skin, it will cause confusion as doctors confuse it with Adult onset Still’s disease when meet a patient who is small intestine Crohn’s disease and has fever only. Corticosteroids are the treatment for Crohn’s disease, and they can also conceal the condition of Crohn’s disease. So if doctors can not find the lesion of a feverish patient, if the patient presents changes of skin and joints, doctors should not diagnose the patient of Adult onset Still’s disease at once, even more, doctors should not use corticosteroids with ease.

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References