Pneumatosis Cystoides Coli Presenting as Acute Abdomen in a Patient with Complicated Behcet’s Disease: A Case Report

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Patient: Male, 40-year-old
Final Diagnosis: Pneumatosis cystoides coli
Symptoms: Abdominal pain • acute abdomen • anorexia
Medication: —
Clinical Procedure: Exploratory laparotomy • ileostomy placement • right hemicolecction
Specialty: Gastroenterology and Hepatology • Rheumatology • Surgery

Objective: Rare disease

Background: Behcet’s disease (BD) is defined as vasculitis involving arteries and veins of any size and affecting almost any organ system. Abdominal manifestations of BD are diverse and nonspecific. Mucosal ulcerations can be seen in the gastrointestinal tract. Extensive ulcerations, especially ileocecal lesions, can lead to perforation, strictures, fistulas, and abscesses. Pneumatosis cystoides intestinale is a rare benign condition characterized by multiple submucosal or subserosal, gas-filled cysts in the gastrointestinal tract wall. Pneumatosis cystoides coli (PCC) affects the colon, can present with a wide range of manifestations, and can mimic many different systemic diseases. We describe a case of PCC in a patient with Behcet’s disease who presented to the Emergency Department with a clinical suspicion of acute abdomen.

Case Report: A 40-year-old man with complicated Behcet’s disease, treated with high-dose steroids, presented with acute abdomen and CT scan findings highly suggestive of intestinal obstruction due to ileocolic intussusception. He underwent laparoscopic right hemicolecction. Pathology demonstrated PCC disease.

Conclusions: Pneumatosis cystoides coli can present with a broad range of symptoms and can be secondary to many systemic and autoimmune diseases. With radiological evidence and a high level of suspicion, unnecessary surgery can be prevented.

Keywords: Behcet Syndrome • Intussusception • Pneumatosis Cystoides Intestinalis

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Background

Pneumatosis cystoides coli (PCC) is an uncommon disease with unclear etiology and pathogenesis [1]. Abdominal pain, diarrhea, and other nonspecific abdominal symptoms are the main clinical manifestations [2]. It is easily confused with intestinal inflammatory bowel disease, polyps, or cancer [3]. The diagnosis mainly depends on abdominal computed tomography (CT) scan and colonoscopy findings [2-4]. The main challenge is to diagnose this entity, especially if the patient has other systemic diseases.

Behcet’s disease (BD) is defined as vasculitis involving arteries and veins of any size and affecting almost any organ system. BD can involve any part of the alimentary tract. Mucosal ulcerations can be seen in the terminal ileum and cecum (most commonly) and along the colon and esophagus. Extensive ulcerations, especially ileocecal lesions, can lead to perforation, strictures, fistulas, and abscesses [5].

We describe a case of PCC in a patient with Behcet’s disease who presented to the Emergency Department with a clinical suspicion of acute abdomen.

Case Report

A 40-year-old man with known complicated Behcet’s Disease (BD) presented to the Emergency Department with abdominal pain and anorexia. He had BD that was diagnosed in 2009 after an episode of aseptic meningitis. His past medical history was also significant for oral and genital aphthae, pericarditis, and myocarditis that caused severe left ventricle dysfunction. Repeated laboratory tests showed negative rheumatological serum tests (antinuclear antibody (Ab), rheumatoid factor, antiphospholipid, and Antiβ2 glycoprotein Ab, anticentromere Ab, DNA Ab, P-ANCA, and C-ANCA), and normal levels of C3 and C4. Genetic tests were not performed. He also had superior vena cava (SVC) obstruction with SVC syndrome manifestations, in addition to pulmonary embolism. He was diagnosed with familial Mediterranean fever (FMF); however, this was not proven genetically. His past surgical history was notable for appendectomy years before he was diagnosed with BD. His regular medications included high-dose prednisone, Imuran, colchicine, beta-blocker (metoprolol), and angiotensin converting enzyme inhibitor, and he was fully anticoagulated with rivaroxaban. He was advised to start anti-TNF treatment but did not complete the pretreatment evaluation.

On physical examination, he had cushingoid habitus, and tenderness and fullness in the right lower abdominal quadrant (RLQ), without peritonitis. His blood tests were within normal limits except for a slightly elevated C-reactive protein (CRP) and subtherapeutic anti-Xa levels. He underwent a CT scan without IV contrast due to a known allergy, which showed a swirl of bowel loops in the right abdomen involving part of the distal ileum, cecum, and ascending colon. Air was noted in the bowel wall, loculated around this swirl (Figures 1, 2). The presentation was not clear and cecal volvulus or ileocecal intussusception were suspected. Due to his concerns and the findings on CT scan, he was taken to the operating room (OR). He underwent a diagnostic laparoscopy, during which a dilated distal small bowel was seen without an obvious transition point. The terminal ileum and cecum were adhered to the abdominal wall, the cecum was folded into itself, small and large bowel seemed viable, and no signs of inflammation like creeping fat or edematous bowel wall were noted. The mesocolon in the area of the cecum was congested and there were some clear small cysts on the adjacent omentum. No obvious internal hernia or intussusception were noted. During the mobilization and adhesiolysis, fullness was felt in the right colon, and a laparoscopic ileocecal resection with side-to-side functional end-to-end stapled ileocolic anastomosis was performed. Although it is not common to create a diverting ileostomy in right colon resection, in this case, due to the high dose of prednisone, severe CHF, and the unclear pathology, we decided to...

Figure 1. Axial image of abdominal CT scan in soft tissue window demonstrating an intussuscept-like swirl of bowel loops (white arrow) in the right lower abdomen involving part of the distal ileum, cecum, and ascending colon.
create a protective ileostomy [6]. The operative and postoperative course were unremarkable. The gross appearance of the specimen showed multiple small soft cysts all over the cecal mucosa (Figure 3). Histological evaluation revealed empty submucosal cysts in the cecum, some of them lined by multinucleated giant cells (Figure 4). No vascular thrombi or signs of vasculitis were observed. The colonic mucosa was negative for atypia, prominent inflammation, cryptitis, crypt abscesses, and granulomas. The morphological picture was consistent with pneumatosis cystoides coli. The loop ileostomy was closed a few months after the first surgery, without complications.

**Discussion**

We present an unusual case of acute abdomen in a patient with complicated Behcet’s disease and pneumatosis cystoides intestinalis and suspicion of intussusception on imaging. This case posed the problem of a broad differential diagnosis that included a complication of his Behcet’s disease, an intussusception, and a benign manifestation of PCC.

**Behcet’s Disease**

BD is defined as vasculitis involving arteries and veins of any size and affecting almost any organ system. The etiology of
BD is unclear and involves an autoimmune process. Behcet’s disease is characterized by oral and genital aphthous, as well as eye involvement (uveitis) and arthritis. Other uncommon possible manifestations include neurological, vascular (arterial aneurysms, stenosis, and venous thrombosis), and cardiac (which can affect any of the layers of the heart as well as the coronary arteries and the heart valves). The clinical picture can mimic many other diseases and the diagnosis is based on clinical symptoms and signs and exclusion of other possible causes. The treatment of BD has evolved in recent years, and the treatment options have been expanded with anti-inflammatory biological drugs.

Abdominal manifestations of BD are diverse and nonspecific. BD can involve any part of the alimentary tract from the esophagus to the colon, including the pancreas (rarely) and the liver. The prevalence of gastrointestinal involvement varies geographically and is more often found in the areas of traditional disease clusters along the ancient “silk road”. Mucosal ulcerations resembling oro-genital aphthae can be seen in the terminal ileum and cecum (most common) and along the colon and esophagus. Extensive ulcerations, especially ileocecal lesions, can lead to perforation, strictures, fistulas, and abscesses. Differentiation from Crohn’s disease is difficult and is based on the overall clinical picture [7].

**Pneumatosis Cystoides Coli**

Pneumatosis cystoides intestinalis is a rare condition characterized by multiple, gas-filled cysts in the gastrointestinal tract wall, found in the submucosa or subserosa. They can occur in the small or large bowel. In the case of colonic cysts, the diagnosis is pneumatosis cystoides coli (PCC). It can be idiopathic (more commonly in the left colon) or secondary (usually in the small bowel and ascending colon) [1]. It is often asymptomatic. When it does become symptomatic, the symptoms can vary and are nonspecific [2]. PCC can also cause almost any secondary abdominal complication, including volvulus, obstruction, hemorrhage, perforation, and intussusception [1,3]. It is usually an incidental finding during surgery or imaging [1]. The unique benign radiographic and endoscopic features are seen in some autoimmune diseases like scleroderma [8], collagen vascular disease, and even Crohn’s disease, but have also been described in COPD patients [1-3]. The pathogenesis of these gas-filled cysts is not entirely clear. There are several theories, but none of them have been proven correct, including pulmonary, bacterial gas formation, and mechanical causes [1-3], in which gas is forced into the bowel wall via several routes, including mucosal break, which can be explained in our patient by vasculitis, connective tissue diseases, and high doses of systemic steroids.

**Conclusion**

We present the case of a complicated Behcet’s disease patient with high clinical and radiological suspicion of acute abdomen due to intussusception. The pathological diagnosis was PCC. The benign finding of PCC was correlated with unspecific symptoms and, although not previously reported, with the patients’ basic complicated BD. The main challenge remains to diagnose this entity and to exclude differential diagnoses, especially if the patient has other systemic diseases.

**Declaration of Figures’ Authenticity**

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**References:**