



Crohn's Disease Manifesting as Acute Appendicitis: Case Report and Review of the Literature

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Abstract

Crohn's Disease (CD) is one of the two clinical presentations of Inflammatory Bowel Disease (IBD) which involves the GI tract from the mouth to the anus, presenting a transmural pattern of inflammation. CD has been described as being a heterogenous disorder with multifactorial etiology. The diagnosis is based on anamnesis, physical examination, laboratory finding, imaging and endoscopic findings. There have been less than 200 cases of Crohn's disease confined to the appendix since it was first described by Meyerding and Bertram in 1953. We present the case of a 24 year old male, who presented with acute onset, right lower quadrant pain, mimicking acute appendicitis with histopathological report of Crohn's disease confined to the appendix.

Introduction

Crohn's Disease (CD) is a chronic entity which clinical diagnosis represents one of the two main presentations of Inflammatory Bowel Disease (IBD), and it occurs throughout the gastrointestinal tract from the mouth to the anus, presenting a transmural pattern of inflammation of the gastrointestinal wall and non-caseating small granulomas. The exact origin of the disease remains unknown, but it has been proposed as an interaction of genetic predisposition, environmental risk factors and immune dysregulation of intestinal microbiota [1,2]. Crohn's Disease (CD) was first described by Dr. Burrill B. Crohn in 1932. It is considered under the spectrum of chronic idiopathic inflammatory bowel disease along with ulcerative colitis.

Dahlhamer et al. recently suggested that approximately 1.3% of the U.S. population is affected by Crohn's disease (roughly 3 million patients), it has an annual incidence that ranges from 3 to 20 cases per 100,000 individuals, with a median onset age of 30 years. There are two described peaks of incidence, the first one between the age of 20 to 30 years and a second smaller one around 50 years [3]. The prevalence of IBD and CD is low in the South Asian region compared with Western countries [4]. In Mexico, a nationwide cohort study reports an incidence of 0.04% per 100,000 persons and a prevalence of 0.34 cases per 100,000 persons [5].

Crohn's disease diagnostic results from correlating clinical signs and symptoms as well as histopathologic endoscopic and laboratory findings. The most common symptoms are chronic diarrhea (>4 weeks), abdominal pain, weight loss and blood or mucous in the stool. There can also be extraintestinal manifestations like primary peripheral arthritis (the most commonly observed), aphthous stomatitis, erythema nodosum and ankylosing spondylitis. The two most common sites of CD are the ileum and the colon [6]. Crohn's disease confined to the appendix is a rare entity, with an incidence that has been reported as 0.1% and up to 2.0% and up to 50% of specimens resected in cases of CD showed appendiceal involvement [2] and was first described by Meyerding and Bertram in 1953. When CD affects the appendix exclusively, it may present with chronic or recurring symptoms, and in most cases it can present macroscopically as an increase in volume that can range from 1.5 cm to 2 cm, adhered to surrounding structures due to chronic inflammatory changes; ultimately, histopathological study will determine whether it is CD or an acute appendicitis non related to CD; additionally appendiceal CD has been considered a different entity from idiopathic granulomatous appendicitis [6]. Appendectomy remains the gold standard for treatment of this disease [7].

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Figure 1: Abdominal CT Scan which revealed mass of appendiceal adhesions, as well as pericecal fat stranding and free fluid which surrounded the caecum, as well as a 26 mm enlarged appendix.

Case Presentation

We present the case of a 24 year old male patient who presented in the emergency department complaining of diffuse abdominal pain, which started 12 h before presenting to the ED with a localization in the upper middle quadrant and afterwards migrated to the right iliac fossa, accompanied by nausea and vomiting, as well as tenderness during deambulation. The patient had no previous medical history. A blood panel revealed leukocytosis ($16 \times 10^3/\mu\text{L}$), bandemia (11%) and neutrophilia (73.2%), no other alterations were revealed on this exam. Upon inspection of the abdomen we observed rebound tenderness and positive Blumberg, Mc-Burney and other appendicular signs. We requested a CT scan with oral contrast which reported a mass of appendiceal adhesions, as well as pericecal fat stranding and free fluid which surrounded the cecum, all of which supported the diagnosis of acute appendicitis.

The patient underwent exploratory laparoscopy, and we observed the presence of a 12 cm appendiceal mass, surrounded by purulent fluid which extended into the pelvic cavity (approximately 60 cc); cultures were obtained for antimicrobial therapy. Trocars were placed in the left iliac fossa, suprapubic space and in the left quadrant. We performed blunt dissection of the mass of appendiceal adhesions, until the appendiceal insertion was found. The appendix was swollen (approximately 15 mm wide) and appeared very erythematous with spots of necrotic tissue which involved the entire appendix; the appendiceal base which was enlarged and with spots of necrosis which extended into the cecum, which also appeared fibrous and erythematous. Given the suspicion of neoplastic disease we decided to perform a laparoscopic ileo-cecal resection with linear stapler (endopath 45 mm Ethicon[®]), and after having transoperative histopathological confirmation that there were no signs of malignant disease we proceeded to reconstruct with an intracorporeal side to side ileo-colonic anastomosis using the same laparoscopic linear stapler and reinforcing with simple vicryl 2-0 sutures. The specimen was extracted carefully with an extra-large endobag by extending the incision in the lower left quadrant, and drainage was placed in the pelvic cavity.

The patient had a normal postoperative period and was started on TPN and with PO liquids on the third post-operative day which was the same day when he passed stools and flatus. The patient required ordinary pain management with NSAIDS and he was discharged 5 days after the surgery.

The definitive histopathological findings reported transmural chronic inflammation, neutrophils, plasmatic cells and eosinophils, transmural lymphoid aggregates, non caseating epithelioid granulomas with giant cells, negative PCR for mycobacterium, consistent with Crohn's disease confined to the appendix, all of



Figure 2: Initial laparoscopic view of cecum and terminal ileum, showing a hardened and enlarged appendix and a mass of appendiceal adhesions.



Figure 3: Swollen appendix adhered to the terminal ileum.



Figure 4: Resection of terminal ileum using linear stapler.



Figure 5: Resection of distal cecum using linear stapler.

which confirmed Crohn's disease confined to the appendix with no apparent cecal involvement.

Discussion

Crohn's disease is considered to be a heterogenous disorder with multifactorial etiology, with a complex interaction between genetics and environment which ultimately cause manifestation of the disease. NOD2, IL23R and ATG16LI genes have been strongly associated to CD. Patients who carry heterogenous NOD2/CARD15 have 2-4 times increased risk of developing CD with ileal involvement, fibrostenotic

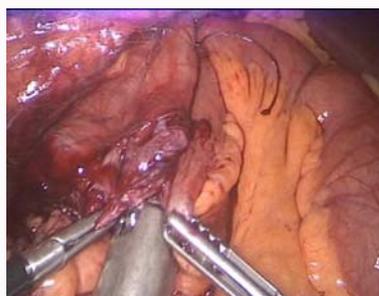


Figure 6: Intracorporeal ileo-colonic side to side anastomosis with linear stapler.



Figure 7: Intracorporeal side to side ileo-colonic anastomosis reinforced with vycril 2.0.

disease and early development of the disease [8]. Environmental risk factors have shown a major impact in CD; smoking has been linked to an influence in phenotype of CD, doubling the risk to present the disease; other risk factors such as symptomatic mumps, high intake of dietary fats and meats, and oral contraceptives have also been associated to CD. Diagnosis is based on thorough physical examination and clinical history as well as imaging, laboratory and endoscopic findings, though there is no single definitive diagnosis tool for CD.

Full ileocolonoscopy with biopsies, demonstrating non-caseating granulomas (present in only 60% of patients) is currently the most widely used diagnostic tool. Panes et al. described ultrasonography to have a higher sensitivity (85% and 98% respectively) and specificity for CD than MRI (78% and 85% respectively) [9]; it also has the advantage of its increased availability and accessibility, lower cost, lack of involvement of ionizing radiation, real time imaging, higher spatial resolution, shorter examination time, no need of sedation or anesthesia, no need for oral or intravenous contrast material [10]. Another systematic review comparing Magnetic Resonance Enterography (MRE) and Computed Tomography (CT), found similar diagnostic results, also without the risk of radiation exposure presented by CT. Capsule endoscopy is a relatively new noninvasive diagnostic technique which has been proven to have a 15% higher diagnostic rate than colonoscopy or ileoscopy [9].

Agha et al. [11] first presented a series of 100 cases of CD presenting as appendicitis, and described a great difficulty in preoperative diagnosis, with narrow appendiceal lumen in barium enema examination, a markedly thickened appendix on sonography or CT, and extrinsic compression of the cecum or terminal ileum suggesting an appendiceal mass.

Appendiceal CD is a rare entity, which has been more frequently described amongst younger patients, ranging from 10 to 45 years

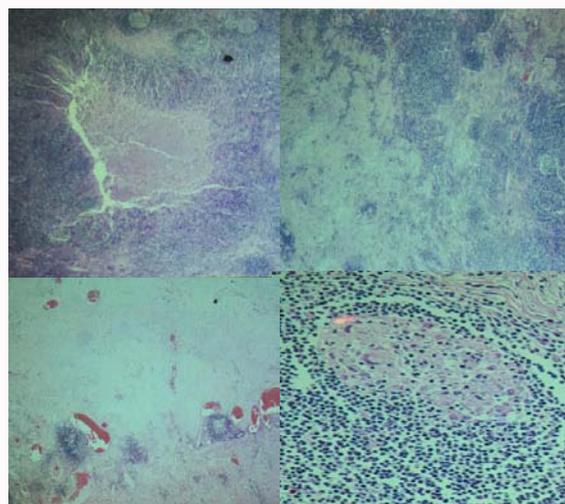


Figure 8: Hystopathological findings demonstrating: chronic transmural inflammation, thickening of the wall with muscularis hypertrophy, neutrophils, plasmatic cells and eosinophils, transmural lymphoid aggregates which spread to the serosa, ulceration and acute inflammation of the mucosa, serosa with lymphoid aggregates, non caseating epitheloid granuloma with giant cells, Negative PCR for mycobacterium.

(average 21.1 years) and predominantly affects male patients. The most common symptom of appendiceal CD is right lower quadrant pain, very similar to that observed in acute appendicitis, which can be acute or chronic (2 or more days). Histopathologic features are characterized by transmural chronic inflammation with marked fibrous thickening of the wall, lymphoid aggregates, small non-caseating granulomas, ulcerative mucosal change, crypt abscesses, muscular hypertrophy, and neural hyperplasia [12]. It has been reported that primary CD of the appendix has a more favorable clinical outcome compared to CD arising in small or large bowel with long post-surgical remissions and a recurrence rate of 8% to 10% [2].

Recently it has been suggested that endothelial Fas-L expression in endothelial cells may play an important role in the regulation of mucosal immunity and in the pathogenesis of CD an IBD [13]. Differential diagnosis should include intestinal tuberculosis, foreign body reaction, and diverticulitis of the appendix, actinomycosis, *yersinia* infection and even carcinoma [14-19]. Ileal resection is clearly indicated for an obstructing ileal stricture that fails to respond to medical or endoscopic therapy, for a right iliac fossa mass with internal or external fistulation, for uncontrolled bleeding, or when there is free perforation [20].

Conclusion

We present a rare presentation of isolated appendiceal Crohn's disease that mimicked as acute appendicitis in a young male. In appendiceal CD the treatment of choice is appendectomy and no further treatment is needed because it has a favorable clinical outcome and low recurrence rate; however there is not clear consensus weather if there's a necessity of follow-up. Some recommend a follow-up of five years. We should have a high index of suspicion to detect recurrences although they are not common to develop. In our patient no recurrence has been noted after follow-up.

An atypical morphological appearance of the appendix should raise suspicion of CD or aetiology. CD when confined to the appendix is less aggressive than when manifests in other sections of the GI tract. It has a low incidence of postoperative fistulae.

The appendix is important for intestinal homeostasis, prevents the development of certain pathologies. Its resection, regardless of whether or not there is an inflammation after surgery, increases the risk of Crohn's Disease and worsens the prognosis of this pathology, so appendectomy should be avoided in the absence of appendicitis.

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