Appendiceal Crohn's disease: a case report

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ABSTRACT

Background: Crohn's disease confined to the appendix is uncommon and is estimated to be 0.2%–1.8% of all appendicectomies.

Case Presentation: Herein, we report a 13-year-old female who presented in the emergency department with typical symptoms and signs of acute appendicitis and elevated inflammatory markers. An abdominal ultrasound was performed which corroborated the initial diagnosis of acute appendicitis. Consequently, the patient underwent an appendicectomy. On operation, the appendix was found to be enlarged, swollen, and with marked thickening of the appendiceal wall. The terminal ileum and the cecum were normal. A drain was placed because of the probable leaking from the appendiceal stamp, which was impossible to bury. Histological findings indicated the diagnosis of the appendiceal Crohn's disease. Postoperatively, the patient recovered without complications.

Conclusion: In a rare situation of appendiceal Crohn's disease, the treatment of choice is appendicectomy and no further treatment is needed.

Keywords: Crohn's disease, acute appendicitis, appendicectomy, histological findings, prognosis.

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Background

Crohn's disease is an idiopathic, chronic inflammatory bowel disorder, which occurs throughout the digestive system from the mouth to the anus, particularly the terminal ileum. The appendix can be affected in about 21% due to the extension from the terminal ileum or the cecum, while isolated Crohn's disease of the appendix is relatively rare with a favorable clinical outcome and low recurrence rate [1]. Nowadays, appendiceal Crohn's disease has become more recognizable and has been considered as a different entity from idiopathic granulomatous appendicitis [2].

Case Presentation

A 13-year-old female visited the emergency department with a high fever (39.2°C), abdominal pain localized in the right iliac fossa the previous 8 hours, with nausea and anorexia. There was no history of diarrhea, constipation, or vomiting. Three days before, the patient developed constant, cramping periumbilical pain and she had visited a local physician, who suggested conservative therapy but she showed no improvement. Abdominal examination disclosed deep tenderness at McBurney point, positive Rovsing's sign, and rebound tenderness in the right iliac fossa. Laboratory studies revealed elevated inflammatory markers (White blood cell count 19.44 K/Ul, neutrophils 89.5%, C–reactive protein 11.80 mg/dl, and erythrocyte sedimentation rate 44 mm/hour). The initial diagnosis was acute appendicitis, which was confirmed by the ultrasonographic findings: enlarged non-compressible blind tubular structure in the right iliac fossa, free fluid around it, and inflammation of the adjacent fat (Figure 1). An appendicectomy was carried out through a McBurney incision. The appendix was identified, exposed, and noticed to be enlarged, hardened, with marked thickening appendiceal wall, but without inflammatory disorder of the

Figure 1. *Abdominal ultrasound: The appendix is enlarged, non compressible, with free fluid around it and inflammation of the adjacent fat.*

cecum and distal ileum (Figure 2). Therefore, a simple appendicectomy was performed. Due to impossible burying of the appendiceal stamp and the prospective leaking from it, a drain was placed. Histopathologic findings evinced the diagnosis of isolated Crohn's disease of the appendix: transmural inflammation with thickening of the appendiceal wall, small non-caseating epithelioid granulomas, lymphoid aggregates, multinucleated giant Langerhan's cells, and muscularis hypertrophy. The drain was removed in the third postoperative day and the patient was discharged in the fifth postoperative day without complications.

Fecal calprotectin after 2 months was 98 μg/mg and the antibodies C–ANCA, P–ANCA, tTG IgA, and ASCA IgG were negative, whereas colonoscopy at the same period showed no evidence of Crohn's disease in the large bowel and the terminal ileum. The patient did not mention any episodes of abdominal pain during her last follow-up visit 3 months after surgery. No further follow-up was recommended in agreement with the gastroenterologist. We review the literature on this rare disorder concerning its prevalence, presentation, diagnostic work-up, and treatment options.

Discussion

Crohn's disease is a chronic inflammatory bowel disease characterized by transmural inflammation, mild disorders of the architecture, and focal distribution of lesions throughout the digestive system. The estimated prevalence of Crohn's disease in the USA is 200 cases per 100,000 among adults and 40 cases per 100,000 among children [2]. The two most common sites are the ileum and the colon. Nevertheless, the appendix could be affected in about 21% due to the extension from the terminal ileum or the cecum [1]. In the original description of the Crohn's disease in 1932, the appendix was not believed to be part of the inflammatory process. Over the next few years, case reports published demonstrating that the appendix could be affected [3]. Isolated Crohn's disease of the appendix was first mentioned by Meyerding and Bertram in 1953 and is being cited with increasing frequency recently [4]. It usually affects young adults in their 20s and 30s and has a male predominance [5]. In our case, the patient was a 13-years-old female, meaning that this phenomenon is not limited to this age group.

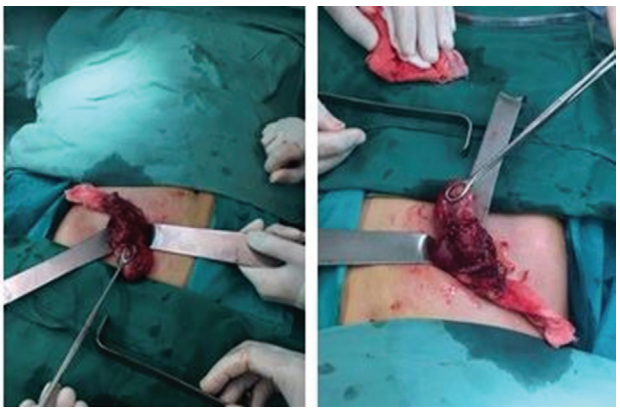

Figure 2. *Surgical specimen: The appendix appears enlarged, swollen, with marked thickening of the wall.*

Initial manifestation of appendiceal Crohn's disease is variable. The most frequent appearance is acute pain in the right iliac fossa suggesting acute appendicitis in about 85% of the patients and chronic pain with a palpable mass in the right iliac fossa in about 25% of patients. Usually, the pain in the right iliac fossa is present for between 3 days and 3 weeks. Other presentations can be a bowel obstruction, intussusception, and rarely lower gastrointestinal bleeding [6]. It is impossible to distinguish Crohn's disease restricted to the appendix from acute appendicitis preoperatively. In contrast, Crohn's disease with appendiceal involvement needs to be distinguished preoperatively from acute appendicitis because it is managed conservatively without surgical operation. Clinical attributes of Crohn's disease are an atypical or protracted clinical course, change in bowel habits, and weight loss [2]. Ultrasonography predictors of Crohn's disease are ileum thickness more than 5 mm and color in the ileum wall with Doppler. Appendix color with Doppler is the only variable significantly associated with the diagnosis of acute appendicitis [7].

Macroscopically, the appendix is markedly enlarged, swollen, indurated, and connected to the periappendiceal soft tissue with fibrous adhesions [8]. Microscopically, the main histological characteristics are transmural inflammation with thickening of the appendiceal wall, small non-caseating epithelioid granulomas, lymphoid aggregates, and mucosal ulceration. Other denotative findings are multinucleated giant Langerhan's cells, crypt abscess, muscularis hypertrophy, neural hyperplasia, and lymphangiectasia [9].

Differential diagnosis includes appendicitis, appendiceal tumors, appendiceal diverticulosis, presence of foreign bodies, and granulomatous diseases of the appendix, including infectious diseases like tuberculosis, actinomycosis, and Yersinia spp, fungal infections such as histoplasmosis and blastomycosis, parasitic infestations, for example, schistosomiasis or *Enterobius vermicularis* [6]. Moreover, appendiceal sarcoidosis may present with granulomas, thick and indurated fibrotic appendix, but it often related with systemic manifestations of the disease [2]. Idiopathic granulomatous appendicitis and appendiceal Crohn's disease are two separate entities. An increased number of granulomas per tissue section is seen in idiopathic granulomatous appendicitis compared to more dispersed granulomas in appendiceal Crohn's disease.

The preferred treatment for appendiceal Crohn's disease is appendicectomy. Appendiceal Crohn's disease is less aggressive and seems to have a much better prognosis than that of Crohn's disease arising in the small or large bowel [9,10]. The postoperative enterocutaneous fistula incidence rate in Crohn's disease limited to appendix has

Table 1. Demographics, clinical presentation, surgical operations, and follow-up of the patients with appendiceal Crohn's disease.

Articles	*P*	Sex	Age	Clinical Presentation	Symptom duration	Clinical Impressions	Surgical Operation	Follow Up
Masuo et al. (1994)	1	M	17	Right Lower Quadrant (RLQ) pain	14 days	Acute appendicitis	Appendicectomy (oedematous appendix)	Barium study and colonoscopy postoperatively, 3 years follow-up
Prieto-Nieto et al. (2001)	10	6 M, 4 F	Average age 29 (range 10–33)	10 RLQ pain, 7 nausea + vomiting, 3 anorexia, 3 fever, 1 diarrhea	1 day–4 months	Acute appendicitis	Appendicectomy (6 oedematous appendix, 3 perforated appendix + periappendiceal abscess, 1 tumoral mass in the appendix)	Mean 14, 5 years (range 2–25)
Han et al. (2014)	12	7 M, 5 F	Average age 29.8 (range 11–51)	9 RLQ pain, 1 abdominal pain, 1 lower abdominal pain, 1 lower abdominal pain + diarrhea	2 days–5 months	10 acute appendicitis, 2 acute appendicitis + perforation	Appendicectomy (10 oedematous appendix, 2 perforated appendix + periappendiceal abscess)	Not mentioned
Lee et al. (2015)	1	F	45	RLQ pain	12 days	Acute appendicitis	Laparoscopically appendicectomy (oedematous appendix)	Colonoscopy preoperatively, capsule endoscopy after 1 month, Magnetic Resonance Imaging (MRI) of the abdomen and pelvis after 3 months
El-Saady et al. (2016)	1	M	24	RLQ pain, nausea, vomiting, anorexia, constipation, fever	3 days	Acute appendicitis	Segmental right hemicolectomy (oedematous appendix)	Colonoscopy after 8 months

Table 2. Histologic features of appendiceal Crohn's disease.

Histologic features	Masuo et al (1994)	Prieto-Nieto et al. (2001)	Han et al. (2014)	Lee et al. (2015)	El-Saady et al. (2016)
Wall thickening	1/1 (100%)	10/10 (100%)	11/12 (92%)	1/1 (100%)	1/1 (100%)
Transmural inflammation	1/1 (100%)	10/10 (100%)	12/12 (100%)	1/1 (100%)	1/1 (100%)
Lymphoid aggregates		4/10 (40%)	12/12 (100%)	1/1 (100%)	1/1 (100%)
Epithelioid granuolmas	1/1 (100%)	8/10 (80%)	12/12 (100%)	1/1 (100%)	1/1 (100%)
Mucosal ulceration		2/10 (20%)	11/12 (92%)		1/1 (100%)
Crypt abscess		2/10 (20%)	5/12 (42%)	1/1 (100%)	1/1 (100%)
Perforation		3/10 (30%)	2/12 (17%)		
Muscular hypertrophy	1/1 (100%)		1/12 (8%)	1/1 (100%)	
Neural hyperplasia			5/12 (42%)		1/1 (100%)
Perpendicular serosal fibrosis		9/10 (90%)	8/12 (67%)		

been reported to be 3.5%, whereas in patients with Crohn's disease of the ileocecal segment rises to 34%–58% [6]. There is a debate as regards the need for follow-up in such patients. Some authors believe that appendicectomy alone is curative in the majority of patients and do not propose surveillance, whereas others recommend follow-up for 5 years [2]. The demographic features, clinical presentation, surgical operations, and follow-up of the patients with appendiceal Crohn's disease are summarized in Table 1, while histopathologic features are summarized in Table 2.

Conclusion

Appendiceal Crohn's disease is a rare condition with an incidence ranging from 0.2% to 0.62% of all appendicectomies. It usually affects young adults in their 20s and 30s and has a male predominance. It is impossible to distinguish appendiceal Crohn's disease from acute appendicitis preoperatively.

Appendiceal Crohn's requires no further treatment after appendicectomy. It seems to have a more benign course compared to Crohn's disease with a reduced rate of complications postoperatively. The importance of follow-up remains controversial. In rare situation of appendiceal Crohn's disease, the treatment of choice is appendicectomy and no further treatment is needed.

Acknowledgment

None.

List of Abbreviations

ASCA IgG	Antibodies against Saccharomyces cerevisiae IgG
C–ANCA	Cytoplasmic antineutrophil cytoplasmic antibodies
P–ANCA	Perinuclear anti-neutrophil cytoplasmic antibodies
tTG IgA	Tissue transglutaminase IgA

Consent for publication

Written informed consent was obtained from the child''s parents prior to publication.

Ethical approval

Not required.

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Summary of the case

Patient (gender, age)	1	Female, 13 year old
Final diagnosis	2	Appendiceal Crohn's disease
Symptoms	3	Fever, pain to the right iliac fossa, nausea, and anorexia
Medications	4	Antibiotics: Cefuroxime, metronidazole
Clinical Procedure	5	Appendicectomy
Specialty	6	General Surgery