

Crohn's disease limited to the appendix, case report

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Received 25 April 2016

Accepted 13 May 2016

The Egyptian Journal of Surgery
2016, 35:460–463

Crohn's disease is a chronic, idiopathic transmural inflammatory disease of the alimentary tract, which may be limited to the appendix in 0.2–1.8% of patients. Here, we report a male patient aged 24 years who presented with typical manifestations of acute appendicitis that proved histologically to be Crohn's disease. On operation, an early phlegmon was found. The appendix was dissected and delivered. It was so large with very thick wall, with induration of the nearby cecal wall. Therefore, segmental right hemicolectomy was performed with ileocolic anastomosis. The specimen was sent for histopathologic examination, which demonstrated Crohn's disease limited to the appendix, with other parts of the specimen free. We reviewed the literature on such clinical entity, its clinical significance, differential diagnosis, and best method of management. Actually, the disease needs to be differentiated from a long list of causes of appendicular granuloma as well as tumors and diverticulosis of the appendix. Histological confirmation is frequently required because of difficulty of conclusive diagnosis during surgery when the disease is limited to the appendix. The disease is usually benign and has indolent course than that developed elsewhere. Some authors debate the need for follow-up at all in those patients, believing that the appendectomy alone is curative in the majority of patients.

Keywords:

appendicitis, crohn's disease, rare inflammatory bowel disease

Egyptian J Surgery 35:460–463

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1110-1121

Case report

A 24-year-old male patient presented with complains of diffuse periumbilical pain 3 days ago, which shifted to the right iliac fossa within 5–6 h. The condition was associated with nausea, vomiting (twice), anorexia, constipation, and fever. Abdominal examination revealed tender Mc-Burny point as well as suprapubic area with rebound tenderness elicited in the right iliac fossa. His pulse was 97 beats/min and temperature was 38.6°C, with unremarkable laboratory values. Ultrasonography demonstrated a large noncompressible tubular blind structure with free fluid around it, mostly acute appendicitis. Subsequently, the patient was prepared for appendectomy. On operation, an early phlegmon was found. The appendix was dissected and delivered (Fig. 1). The wall was so thick (Fig. 2), which led to debate about the safety of simple appendectomy, especially with the feeling of cecal induration (shown in Fig. 3). Therefore, segmental right hemicolectomy was performed with ileocolic anastomosis (Fig. 4). The specimen was sent for histopathologic examination, which demonstrated Crohn's disease of the appendix with nonspecific typhlitis in the nearby cecum. The major remaining part of the cecum, the part of ileum, and the ascending colon in the specimen were free. Histopathologic revision of the slide was carried out with confirmation of isolated Crohn's disease of the

appendix. The patient did well in the postoperative period. Colonoscopic assessment was carried out after 8 months with normal colonoscopic study. We reviewed the literature on such clinical entity, its clinical significance, and best method of management.

Discussion

Crohn's disease is a chronic, idiopathic transmural inflammatory disease with a propensity to affect the distal ileum, although any part of the alimentary tract can be involved [1]. A prevalence of about 200 cases per 100 000 has been suggested recently for Crohn's disease in the USA [2]. In the original description of Crohn's disease, the appendix was not believed to be involved in the inflammatory process. Later on, case reports started to appear in publications, demonstrating that the appendix could be involved in the inflammatory changes of Crohn's disease, and it could also be the primary or the sole manifestation of the disease [3]. Isolated involvement of the appendix in Crohn's disease is reported to be 0.2–1.8% and is usually associated with ileocaecal Crohn's disease in

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Figure 1



A large appendix was delivered after its dissection from early phlegmon.

Figure 2

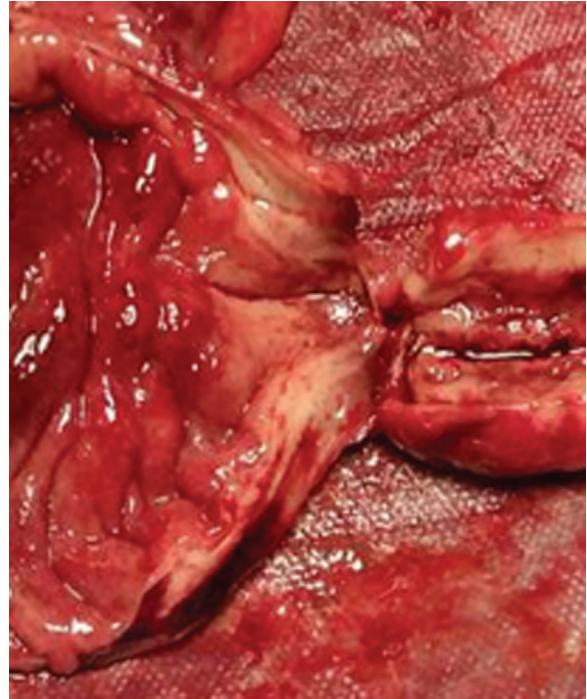


The involved appendix opened longitudinally demonstrating submucosal affection of its whole length.

25% of ileal and 50% of cecal disease [4]. Crohn's disease limited to the appendix usually affects young adults in their 20s and 30s, although it is not limited to this age group and can occur at any age [5].

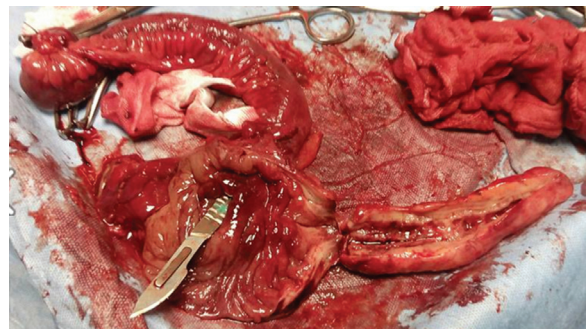
The pathologic features are identical to the disease elsewhere [6]. Macroscopically, the appendix is enlarged, swollen, and adherent to the surrounding structures, these findings being secondary to chronic inflammation. Histologically, the disease is characterized by transmural inflammation with thickening of the appendiceal wall, epithelioid granulomas, lymphoid aggregates, and mucosal ulceration.

Figure 3



Cecal involvement of this thickening.

Figure 4



The specimen of right hemicolectomy in this case.

Other histological features are multinucleated giant Langherhan's cells, crypt abscess, neural hyperplasia, and lymphangiectasia [7].

The clinical presentation of appendiceal Crohn's disease entity is variable. A picture of acute appendicitis with acute pain in the right iliac fossa occurs in about 85% of patients, and about 25% of patients presented with chronic abdominal pain and a palpable mass in the right iliac fossa [4]. Bowel obstruction can be another acute presentation of Crohn's disease of the appendix [4]. Intussusception can be predisposed by the diseased appendix [8]. Complete appendicular inversion, the 'inside-out' appendix with intussusception of the appendix into

the cecum, had also been reported [9]. Occasionally, lower gastrointestinal bleeding from appendiceal hemorrhage had been recently reported and may be the initial or main presentation [10,11]. When the clinical presentation and preoperative diagnosis is acute appendicitis, Crohn's disease may be suspected when there is previous change in bowel habits or an atypical or protracted clinical course is present in a patient before the appendicitis, particularly in areas where Crohn's disease is prevalent [4].

The disease needs to be differentiated from different appendicular pathologies such as appendicular tumors [12], appendicolith [13], diverticulosis of the appendix [14], and granulomatous diseases of the appendix [15]. The latter have several causes, which may be infectious agents such as tuberculosis, actinomycosis, or *Yersinia* spp. [16] and rarely fungal infection such as histoplasmosis or blastomycosis, or a parasitic infestation such as schistosomiasis or *Enterobius vermicularis* [15]. Moreover, appendiceal sarcoidosis, which is rare, may present with granuloma with thick, indurated fibrotic appendix, but it often associated with systemic manifestations of the disease [4]. Granulomatous inflammatory disease isolated to the appendix is similar to Crohn's disease of the appendix but needs to be differentiated when its long-term course in the majority of patients is extremely benign [15]. It differs from typical Crohn's disease of the appendix by less incidence of the development of enterocutaneous fistulas and recurrence [15]. Some believe that isolated granulomatous appendicitis without small bowel or cecal involvement may not represent true Crohn's disease [15]. However, isolated granulomatous appendicitis had been reported to progress to fulminant Crohn's colitis shortly after appendicectomy [11]. Preoperative diagnosis of the disease is actually difficult, especially in acute presentation. However, some tools may be helpful. Ultrasonography and color Doppler sonography may be useful for differentiating Crohn's disease with appendicular involvement from acute appendicitis [17,18]. Computed tomography may be helpful in the diagnosis [19]. It can demonstrate increased appendiceal wall enhancement at a statistically significant level. This is frequently observed in patients with active Crohn's disease [20]. The use of technetium-tagged white blood cells may be helpful in some cases [21]. When the disease is located atypically in the appendix, histological evaluation is required as during surgery a conclusive diagnosis is difficult [3].

The diagnostic difficulty arises when a patient undergoes an appendicectomy for suspected acute appendicitis and

the surgeon unexpectedly encounters ileocaecal pathology whose nature is difficult to ascertain in an emergency situation. In view of this, a definitive treatment in the form of ileocaecal resection for conditions like Crohn's disease may be difficult to carry out. Frozen section, although it may be helpful to differentiate some of these conditions, may usually not be feasible in an emergency situation [4].

If Crohn's disease of the appendix is suspected and is limited to the appendix then appendicectomy alone is a sufficient surgical procedure with very low intraoperative and postoperative mortality and a low rate of fistula formation [5]. The incidence rate of postoperative enterocutaneous fistula in Crohn's disease restricted to the appendix alone has been reported to be 3.5%, whereas in patients with Crohn's disease extended to the ileocaecal segment the postoperative enterocutaneous rate rises to 34–58% [5].

It appears that the course of Crohn's disease limited to the appendix is more indolent than Crohn's disease of the ileum or colon, with a recurrence rate in the largest series of 8%. The interval time from the diagnosis until the recurrence varied from 1 to 48 months, with an average of 19 months. Some authors debate about the need for follow-up in those patients, believing that the appendectomy alone is curative in the majority of patients, whereas others recommend follow-up for 5 years [3].

Conclusion

Crohn's disease is a chronic, idiopathic transmural inflammatory disease of the alimentary tract, which may be limited to the appendix in 0.2–1.8% of patients. Such entity usually presented acutely as acute appendicitis, intestinal obstruction, or lower gastrointestinal bleeding, although some cases presented with chronic abdominal pain and or mass. The disease needs to be differentiated from different appendicular pathologies such as appendicular tumors, appendicolith, diverticulosis of the appendix, and granulomatous diseases of the appendix. When the disease is located atypically in the appendix, histological evaluation is required, as during surgery a conclusive diagnosis is difficult. The disease limited to the appendix is usually benign and has indolent course than that developed elsewhere. Some authors debate the need for follow-up at all in those patients, believing that the appendectomy alone is curative in the majority of patients. Others recommend follow-up for up to 5 years [3].

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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