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Case Report

ACUTE APPENDICITIS REMAINS A GREAT MIMICKER – THE PITFALLS IN THE DIFFERENTIAL DIAGNOSIS AND TACTICS - A CASE REPORT

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Summary

Acute appendicitis (AA) is the most common non-traumatic abdominal emergency. Despite the improved knowledge, experience, and technological advance, its diagnosis remains a challenge. Herein we report an example of a difficult diagnosis of acute appendicitis and comment on the possible pitfalls in the differential diagnosis and surgical tactics. We present the case of a 41-year-old man who had been admitted to another hospital with an initial diagnosis of acute appendicitis and changed to Crohn's disease (CD). Because of a pelvic abscess, percutaneous drainage had been performed. Thrombosis of the right femoral vein had been diagnosed and treated accordingly. In an improved condition, he was referred for elective operation with a final diagnosis of neuroendocrine tumour based on cytology. At laparotomy, the appendix was found densely adherent to the right external iliac vein with a well-demarcated tumour (1 cm) at the base. Appendectomy with partial resection of the caecum with a linear stapler was performed. The histological examination revealed acute to chronic appendicitis with lymphoid follicle hyperplasia at the base. The case illustrates the necessity for broad differential diagnosis in AA and the possibility of severe vascular complications in complicated AA. Taking a detailed history and CT are of paramount importance for an accurate preoperative diagnosis, especially of CD. All emergency surgeons should also be familiar with the scenario of unexpected findings at laparotomy, especially with the management of CD and the algorithms for treatment of appendiceal malignancies. The mini-invasive drainage of right iliac fossa abscess allows for optimizing the patient's condition and may help to avoid unnecessary extensive resections.

Keywords: acute appendicitis, differential diagnosis, Crohn's disease, appendiceal malignancy.

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Introduction

The incidence of acute appendicitis (AA) is about 90-100/100 000 per year, but a sizeable geographic difference has been reported with the lowest rate in Africa [1]. It is the most common non-traumatic abdominal emergency. Despite the improved knowledge and technological advance, the diagnosis of AA remains a challenge [1].

We report an example of a difficult diagnosis of

acute appendicitis and comment on the possible pitfalls in the differential diagnosis (DD) and surgical tactics.

Case report

A 41-year-old man, diagnosed with a neuroendocrine tumour of the appendix, was referred to our clinic after a 20-day stay in another hospital. He had been there on account of right iliac fossa pain, nausea, vomiting, intermittent diarrhea, and temperature up to 38° C. On admission, results from the blood tests were normal except for leukocytosis (15x10^9/L) and CRP of 54 nmol/L. The abdominal examination revealed a palpable mass with moderate pain in the right lower quadrant without rebound tenderness and normal bowel sounds. On the initial ultrasound examination, AA was suspected. The subsequent computed tomography suggested Crohn's disease (CD) – dilated terminal ileum with thickened and contrast-enhancing wall tightly adherent to the caecum and sigmoid colon, and suspected appendicitis in the middle of the formation (Figures 1, 2).

Antibiotic treatment with broad-spectrum antibiotics was then started. Colonoscopy revealed a tumour-like structure with sized 2-3 cm, located in the opening of the appendix and the otherwise healthy colon and terminal ileum. A CT scan performed five days later because of the lack of improvement, found a sizeable pelvic abscess (Figure 3).

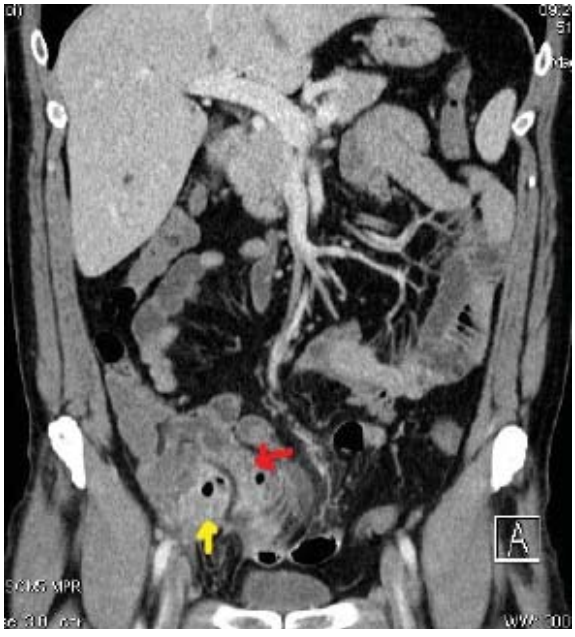

Figure 1. The terminal ileum with a thickened and contrast-enhancing wall (red arrow) and a suspected appendix surrounded by inflammatory mass tightly adherent to the cecum (yellow arrow)

Figure 2. The appendix adjacent to the right iliac vessels (red arrow) and close to a dilated up to 5 cm ileal loop (yellow arrow) adherent to the middle third of the spastic sigma (green arrow)

Percutaneous drainage of the abscess cavity with 9 Fr pigtail catheter was performed. The cytologic examination of the lavage fluid raised suspicion for neuroendocrine tumours. On the 15th day, the patient complained of pain and an increased circumference of the right lower leg.

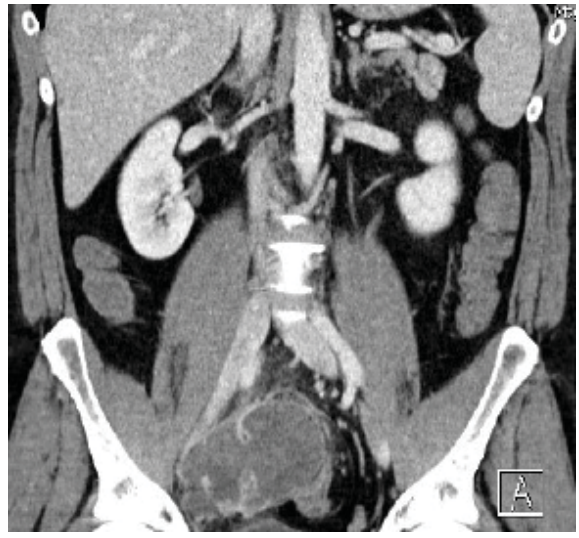

Figure 3. Right pelvic abscess abutting the right external iliac vein

He was consulted with a vascular surgeon. The Doppler examination revealed thrombosis of the right external iliac and femoral vein, which was successfully treated with Nadroparine calcium. After improvement, the patient was discharged and referred for elective operation with a final diagnosis "neuroendocrine tumour of the appendix".

On admission to our clinic, the patient had normal abdominal status and laboratory values. At the midline laparotomy, a massively adherent conglomerate between the distal ileum, appendix, and the middle third of mesosigma was found. The appendix was 5 cm in length with a thickened wall and densely adherent to the right external iliac vein. A tumour-like formation with sized 1 cm at the base was found. Appendectomy with partial resection of the caecum with a linear stapler was performed. Grossly, a yellowish and well-demarcated tumour about 1 cm in size was found at the base, with complete obstruction of the lumen. The histological examination revealed signs of chronic appendicitis with lymphoid follicle hyperplasia at the base and pronounced periappendicular fibrosis. The patient had an uneventful recovery.

Discussion

The differential diagnosis (DD) of AA usually includes a wide range of gastrointestinal diseases: CD, diverticulitis, mesenteric adenitis, infectious enterocolitis, tuberculosis, as well as various genitourinary and gynaecological pathologies [2]. The diagnostic process is additionally complicated by the multiple possible anatomic locations of the appendix and the lack of pathognomonic signs. Various scoring systems have been proposed in the literature, but none of them is highly specific [1]. Herein we focus on only two possible DD scenarios due to the frequently difficult tactical decisions – CD and malignancies of the appendix, which are encountered at laparotomy for AA.

The initial manifestation of CD may mimic AA with three possible scenarios: isolated CD of the appendix, acute appendicitis in known CD, and newly diagnosed CD with a normal appendix at laparotomy.

Isolated appendiceal CD (aCD) is rare, with about 253 cases described in the literature up to 2017 [3]. The clue to the right diagnosis is taking a detailed past medical history. In contrast to the typical AA, aCD is characterized by prolonged complaints of right lower abdomen pain, in some instances years before admission. In one-third of the cases, aCD manifests with diarrhea, containing mucus and blood and palpable mass [4]. Appendectomy is usually the definitive treatment, and this localization is considered as a less aggressive form of the disease. The second possible scenario is AA in known, ileocecal CD (also rare - 3% of all CD) [5]. The dilemma is whether to perform simple appendectomy or ileocecal resection. Although appendectomy can be a definitive treatment, the complications can reach 10-30%, so many authors advocate ileocecal resection in these cases [6-8].

The most challenging scenario is a newly diagnosed CD with a normal appendix at laparotomy, because the appendectomy is associated with complications in one-third of these cases [9-11]. The most recent CD consensus does not recommend resection for terminal ileitis without penetration or obstruction [12]. On the other hand, a 13-year-follow-up of 36 cases revealed a 50% rate of second operative intervention after initial ileocecal resection versus 92% in cases, when only appendectomy was performed [13]. In routine practice, due to the lack of enough experience, this problem is frequently left at the discretion of the operating surgeon.

The modern imaging tools help surgeons in the decision-making process. In AA, a CT scan has 92-98% sensitivity and 100% specificity [1, 2, 14]. CT also has the advantage to discriminate AA from CD. The characteristic features of CD are the thickening and marked stratification of the bowel wall, "the comb sign," and "the fat creeping sign" [2, 5].

The second major problem requiring a sound and adequate intraoperative decision includes the primary malignancies of the appendix with a rate estimated as 0.12 per 1 million people per year [15]. According to an analysis of a large cohort with 2514 malignancies, the most common tumour types were mucinous cancer (38%), followed by adenocarcinoma (26%), and neuroendocrine tumour (17%) [16]. The most common type with a size of less than 1 cm was that of goblet tumour, whereas the

mucinous tumours most frequently were larger than 2 cm. Of note, inappropriate operative procedure (simple appendectomy) was performed in 30% of noncarcinoids and in 28% of the neuroendocrine tumours larger than 2 cm. Based on such extensive experience, the authors recommend a treatment algorithm, according to which all noncarcinoids should undergo right hemicolectomy, as well as for carcinoids > 2 cm or less, but localized at the base of the appendix.

A right iliac fossa abscess can occur in a variety of abdominal diseases – acute appendicitis, CD, cancer, among others. A periappendicular abscess may occur in up to 4% of the cases with AA [17]. The non-drained abscesses have been more frequently associated with right hemicolectomy. In CD complicated with abscess, the National survey in the USA reported an increased rate of percutaneous drainage from 7% in 1998 to 29% in 2007 [18]. The literature results, however, are conflicting. The Cleveland Clinic reported control of the sepsis with subsequent elective operation in 65% of the cases and lower hospital costs [19]. Others, however, have reported prolonged hospital stay and elective intervention in only 9% [20, 21]. A large meta-analysis showed a seven-fold increased risk for recurrence and avoidance of surgical intervention in only 30% [21]. Despite the heterogeneity of the literature, we consider percutaneous drainage as a useful initial step for optimizing a patient's condition through nutrition and sepsis control. It can be definitive or allow for avoiding extensive resection or stoma and achieving lower postoperative morbidity, such as in the case presented here [22-25].

Last but not least, laparoscopy could be useful in both diagnostic and curative approaches despite the higher rate of conversion in males, as well as in emergency settings [26-28].

Conclusion

The case we present illustrates the necessity for broad differential diagnosis in AA and the possibility of severe vascular complications in complicated AA. Taking a detailed past history and CT are of paramount importance for the exact preoperative diagnosis, especially of CD.

All emergency surgeons should also be familiar with the scenario of "unexpected finding" at laparotomy, especially with the management of CD and the algorithms for treatment of appendiceal malignancies. The mini-invasive drainage of right iliac fossa abscess allows for optimization of the patient's condition and may help to avoid unnecessary extensive resections.

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