Incidental Appendiceal Diverticulosis Associated with Acute Appendicitis: A Case Report and Review of Literature

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Abstract

Background: Appendiceal diverticulum (AD) is a rare surgical finding. This entity is classified as either congenital or acquired, with a reported incidence of 0.014% and 0.2-1.7%, respectively. AD can often be associated with acute or chronic appendicitis. Clinically, AD must be identified before any surgical procedure as its concomitant presence with acute or chronic appendicitis increases the risk of perforation and may be associated with underlying neoplasia such as low-grade mucinous adenoma. We hereby report a case of incidental appendiceal diverticulosis associated with acute appendicitis in an attempt to increase the physicians’ perception, knowledge, and awareness of this rare medical condition.

Case Presentation: We report a 40-year-old male patient, with no relevant family history, who presented to the Emergency Department after four days of vague and moderate epigastric pain and nausea. The physical examination upon presentation was unremarkable. Computed Tomography (CT) scan was done and was suggestive of an inflamed appendix. The patient underwent an urgent laparoscopic appendectomy. The removed appendix was inflamed and grossly abnormal with multiple diverticulae. The patient had a smooth recovery.

Conclusion: Diverticulosis of the appendix is a rare clinical entity and, as in our case, is often diagnosed during or after appendectomy. When associated with acute appendicitis, it may be suspected preoperatively with the appropriate imaging technique. This is of paramount importance as it can rapidly progress to perforation and/or lead to a higher mortality rate.

Keywords: Acute appendicitis; Appendix; Appendiceal Diverticulosis; Rare; Case Report
Introduction

Diverticular disease of the appendix is a rare entity and is identified in only 0.004-2.1% of surgical appendectomy specimens and in 0.20-0.66% of routine autopsy samples [1, 2].

It was first described by Kelynack in 1893, with only few publications related to this disease till this date [3]. Appendiceal diverticulum (AD) is classified into two categories: congenital (true) and acquired (false), with the latter being much more prevalent (0.014% for the congenital versus 0.20 to 1.7% for the acquired) [4].

The acquired type occurs on the mesenteric border of the appendix and is usually associated with an arteriolar vessel [5]. Male sex, older age and history of Hirschsprung’s disease and cystic fibrosis have been reported as risk factors for the development of appendiceal diverticulosis [6].

In the context of acute appendicitis, the diagnosis of incidental AD occurs largely intra- or post-operatively as the diverticula have limited clinical characterization and are commonly overlooked by radiologists during the pre-clinical workup [7]. Nonetheless, it is crucial to diagnose this entity pre-operatively as it is associated with a four-fold increase in the risk of perforation as compared to simple acute appendicitis.

In addition, it carries significant association with underlying concomitant malignancy [8]. Indeed, Dupre et al. reported concomitant appendiceal neoplasm such as carcinoids, mucinous adenomas, tubular adenomas and adenocarcinomas in up to 48% of the analyzed specimens. Similar findings were reported by Lamp et al., whereby low grade appendiceal mucinous neoplasms were observed in 42% of the specimens [9,10].

Whether associated with acute appendicitis or not, the treatment of choice for AD is appendectomy [6]. We hereby report a case of incidental appendiceal diverticulosis associated with acute appendicitis in order to increase the physicians’ perception, knowledge, and awareness of this rare medical condition.

Case Presentation

A 40-year-old male patient with a history of left renal cell carcinoma managed by laparoscopic partial nephrectomy, presented to the emergency department with abdominal pain that started four days prior to presentation. The patient described the pain as vague and epigastric, and reported nausea. The pain was increasing in intensity with time, was not related to any food intake, was only transiently relieved by pain killers, and aggravated by physical exercise and cough. In addition, the patient denied any history of a recent weight loss, altered bowel habit, or urinary symptoms.

Upon presentation, the patient was hemodynamically stable. He was afebrile, normotensive, with a normal heart rate and
respiratory rate. The physical exam was unremarkable. The abdomen was soft with positive bowel sounds and no rebound tenderness, peritoneal signs, or muscular rigidity. Murphy’s sign was negative. All laboratory studies were within the normal ranges including pancreatic enzymes (amylase, lipase), and liver enzymes (alanine and aspartate aminotransferases ALT and AST, alkaline phosphatase, ALP, and gamma-glutamyl transpeptidase GGT) panels. A Computed Tomography (CT) scan of the abdomen and pelvis was performed and revealed lymphoid hyperplasia and an enlarged appendix measuring 1.1 cm in diameter with two appendicoliths, along with a prominent fluid-filled tip associated with minimal surrounding fat stranding, all of which were suggestive of tip appendicitis (Figure 1). Therefore, an urgent laparoscopic appendectomy was performed.

Grossly the appendix was dilated with three abnormal small thick-walled lesions (diverticula) identified at its tip (Figure 2). The mesoappendix was eventually excised using ligature and the appendix was removed in an endopouch through the umbilicus.

Macroscopic examination revealed multiple diverticula at the tip of the appendix, measuring between 0.3 to 0.8 cm, along with the presence of few fecaliths. Microscopic examination revealed a non-inflamed false (pseudo) diverticulum on a background of acute appendicitis as demonstrated by a transparietal neutrophilic inflammatory infiltrate associated with focal mucosal erosion and prominent vascular congestion (Figure 3). The patient’s postoperative course was uneventful. He was discharged home on the first postoperative day.

Discussion

Appendiceal diverticula are histologically divided into two main categories: true and false, with the latter accounting for the majority of cases including the one being reported here [1]. AD are further classified into four different subtypes based on the combined healthy and/or pathological state of the diverticulum and appendix, respectively [11]. Type 1 AD (classic type, most common) includes a normal-appearing appendix with acute diverticulitis [1]. Type 2 features an acute diverticulitis accompanied by appendicitis. Type 3 corresponds to a conventional appendicitis with an incidental finding of uninfamed appendiceal diverticulum, similar to our case. Finally, Type 4 AD comprises an incidental non-infamed diverticulum with non-infamed appendix [1,5,11].

Figure 2: Gross appearance of the appendix in vivo with evidence of multiple abnormal thick-walled lesions (diverticula) on its tip (arrow).

Macrocopistic examination revealed multiple diverticula at the tip of the appendix, measuring between 0.3 to 0.8 cm, along with the presence of few fecaliths. Microscopic examination revealed a non-infamed false (pseudo) diverticulum on a background of acute appendicitis as demonstrated by a transparietal neutrophilic inflammatory infiltrate associated with focal mucosal erosion and prominent vascular congestion (Figure 3). The patient’s postoperative course was uneventful. He was discharged home on the first postoperative day.

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Figure 3: Histological section of the appendix and its diverticulum showing a non-infamed false diverticulum on a background of acute appendicitis as demonstrated by a transparietal inflammatory exudate rich in neutrophils and associated with focal mucosal erosion and prominent vascular congestion. D: diverticular lumen; A: Appendicular lumen.

Congenital diverticula are true diverticula because they contain all the layers of the appendiceal wall, including the mucosa, submucosa, serosa, and most importantly, the muscle layer which is usually hypertrophied [1, 12]. Acquired diverticula are false diverticula as only the mucosa, submucosa, and serosa contribute to the wall of the diverticulum without the muscle layer [1, 4, 5, 8, 12]. The latter occurs by herniation through a defect in the
muscularis propria in the mesenteric area at the site of the penetrating arteries [4, 8, 11, 12]. This categorization is important because the muscular layer confers greater resistance to perforation in the setting of a congenital diverticula [1]. In our case, the etiology of the diverticula is probably linked to a sustained contraction of the appendix upstream to two obstructing appendicoliths with subsequent luminal distension and concomitant inflammation.

Acquired appendiceal diverticulosis tend to occur in older adults (>30 years old) with a male sex predilection [1, 7]. AD has many mimickers including acute appendicitis, Crohn’s disease, intestinal tuberculosis, intestinal parasitic infections, appendiceal neoplasm (in case of large diverticula associated with mass effect as a result of abscess or cyst formation), and gynecological diseases including tubo-ovarian abscess, pelvic inflammatory disease, and endometriosis [1]. Appendiceal diverticulosis is usually asymptomatic and diagnosed post-operatively in most of the cases [1]. In a study conducted on 25 cases of AD in a Kuwaiti hospital between 2003 and 2011, none of the cases were diagnosed intra-operatively or even pre-operatively by imaging studies [7]. In another series by Deng YW et al., ten cases of AD were reviewed and all of them were diagnosed after appendectomy [6].

Pre-operative diagnostic studies for AD includes both ultrasound and CT scan examination, with the latter being most accurate for pre-operative diagnosis [7, 8, 12]. However, their diagnostic yield is influenced by the radiologist’s experience, and AD is frequently overlooked [8,12]. Laparoscopic exploration offers both a diagnostic and therapeutic perspective. It remains the sole therapeutic approach whenever AD is suspected to prevent adverse complications such as perforation and neoplastic degeneration [12]. In fact, two-third of cases with uninfamed AD develop acute or chronic diverticulitis, which in turn can proceed to perforation probably due to lack of a muscular layer within the diverticula [1]. It is documented that the rate of perforation of appendicitis with AD (27%) is higher than that of appendicitis alone (6.6%) [13]. Perforation is usually retroperitoneal and leads to localized peritonitis [2]. Other serious complications include gastrointestinal tract hemorrhage requiring blood transfusion, abscess formation, development of appendico-vesical fistula and pseudomyxoma peritonei [1].

Conclusion

We presented a rare case of acute appendicitis with an incidental finding of appendiceal diverticulosis. This diagnosis remains challenging to radiologists and clinicians as they are still, to a high extent, unaware of its clinical features. The radiologist familiarity and experience with this clinical entity is crucial for pre-operative diagnostic imaging. Diagnostic algorithms that include clinical, radiological and pathological features are needed to enhance our diagnostic yield and treat AD accordingly.

References

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