CASE REPORTS
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ADENOCARCINOMA OF THE APPENDIX MIMICKING COMPLICATED APPENDICITIS IN THE ELDERLY – A REPORT OF TWO CASES

ADENOKARCINOM APENDIKSA KOJI IMITIRA KOMPLIKOVANI APENDICITIS KOD STARJIH PACIJENATA – PRIKAZ DVA SLUČAJA

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Summary
Introduction. Primary appendiceal adenocarcinoma is a very rare malignancy which accounts for 0.1% of all appendectomy specimens. In both patients presented in this paper, appendectomy was performed due to suspected acute complicated appendicitis. Case Reports. The first patient, a 77-year-old man, presented with a low grade colonic-type pT3 adenocarcinoma of the appendix, diagnosed by histopathological examination of the resected appendix delivered in a fixative. A month after appendectomy, the patient underwent right hemicolecctomy of a tumor at the edge of the resection. Due to a cardiovascular disease, adjuvant chemotherapy was not indicated. The second patient, a 74-year-old female, presented with a low grade mucinous adenocarcinoma of the appendix with subserous infiltration, diagnosed by histopathological analysis of the resected appendix. Eight months after appendectomy, the patient developed a recurrent tumor in the cecal area. After radical surgical excision of the recurrent tumor, the patient received adjuvant chemotherapy. Both patients had a 5-year survival without relapse. Conclusion. Preoperative diagnosis of appendiceal adenocarcinoma is a challenge due to overlapping symptoms of complicated acute appendicitis. Our results suggest that in elderly patients with symptoms of complicated acute appendicitis, appendectomy should be done with intraoperative histopathological frozen section consultation. In advanced stages of adenocarcinoma, right hemicolecctomy is a better choice than appendectomy.

Key words: Adenocarcinoma; Appendiceal Neoplasms; Appendectomy; Diagnosis; Acute Disease; Appendicitis; Morphological and Microscopic Findings

Sažetak

Ključne reči: adenokarcinom; neoplazme apendiksa; apendektomija; dijagnoza; akutna bolest; appendicitis; morfološki i mikroskopski nalazi

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**Introduction**

Primary appendiceal adenocarcinoma (AA) is a rare malignancy with an incidence of 0.12 cases per 1,000,000 persons per year [1]. Primary neoplasms of the appendix are histologically diverse. They are broadly classified as colonic-type adenocarcinoma, mucinous adenocarcinoma, goblet cell adenocarcinoma, or neuroendocrine carcinoma [2]. Carcinoids are the most common tumors of the appendix (66%), followed by mucinous adenocarcinoma (20%), and colonic type adenocarcinoma (10%) [3]. Most primary tumors of the appendix occur in the elderly, between the fifth and seventh decades of life, with the exception of carcinoids, that are usually diagnosed before the age of 40 [3]. The clinical symptoms of the tumors of the appendix are nonspecific and resemble the symptoms of acute appendicitis, rarely to other gastrointestinal or urogenital diseases [4]. The five-year survival rate for AA is 49 - 60%, and the prognosis is favorable when the tumor is diagnosed at an early stage of the disease [4]. We present two cases of AA which were clinically mimicking complicated appendicitis and manifested with symptoms of acute abdomen.

**Case 1**

A 77-year-old male was admitted to our hospital with a right lower quadrant abdominal pain and peritoneal tenderness. The patient complained about melena, and constipation lasting a few days. The patient’s medical history showed age-related cardiovascular disorders. Routine laboratory test results were within normal limits. Ultrasound examination showed a dilated and thickened appendix, hypoechoic with loss of normal mural stratification, and appendicular lump with free fluid in the peritoneal cavity. Laparotomy and appendectomy were performed. On gross section, the proximal part of the resected appendix was firm, dilated and ruptured with purulent exudate on the surface. Histologically, an invasive colonic type, low grade adenocarcinoma of the appendix (Figure 1), infiltrating up to subserosa was noted. There was no lymphatic or vascular invasion. The right hemicolecction was done a month after appendectomy, and the histologic examination revealed that the rest of the tumor invaded the cecal wall up to the serosal surface. Metastases were not found in any of the 15 ileocecal lymph nodes. The postoperative course was uneventful. Adjuvant chemotherapy was not applied due to cardiovascular comorbidity. The patient was disease-free 62 months after the surgery, which was determined by imaging techniques.

**Case 2**

A 74-year-old female was admitted to our hospital due to abdominal pain and peritoneal tenderness. The patient complained about constipation, bloating, and loss of appetite in the last few months. She was subfebrile (37.7°C) during the last month, especially in the evening. A complete blood count revealed only an elevated white blood cell (WBC) count: 10.5 x 10^9/l. Tumor markers were within the normal range. Ultrason sound examination showed an appendicular lump with a cystic space and free fluid in the peritoneal cavity. A laparotomy with appendectomy was performed. On gross section, the resected appendix was dilated and ruptured. It was softened with mucous content on the surface and impacted mucus in the wall. The histopathological diagnosis was low grade mucinous adenocarcinoma of the appendix without lymphvascular invasion (Figure 2). The postoperative course was uneventful and the patient did not respond to the calls for consideration of the Oncology Advisory Board. Eight months after appendectomy, the patient was readmitted due to abdominal pain, constipation, and poor appetite. She lost 10 kilos in the last seven months. Colonoscopy revealed a tumor in the cecal region and right hemicolectomy was done. The tumor was diagnosed as a recurrent cecal invasive mucinous adenocarcinoma staged as r T3 N0 Mx. The patient underwent adjuvant therapy with Capecitabine, 1,250 mg/m^2/day for 14 days every 21-days, for a total of 6 cycles. The patient was disease-free at follow up, 65 months after the second surgery.

**Discussion**

During the 21st century, the pooled incidence of appendicitis was (in per 100,000 person-years): 100 for Northern America; 113 for Northern Europe; 112 for Southern Europe; 105 for Eastern Europe, and 151 for Western Europe [5]. Appendicitis is a common disease in the elderly, accounting for 5% of all abdominal surgical emergencies in this population [6]. Classical presentation of appendicitis with
abdominal pain showing typical migration from epigastrium to the right lower abdominal quadrant and mild WBC elevation may not be representative in this subgroup of patients. Appendicitis in the elderly needs special attention, because atypical presentations and complications are common [7]. The laparoscopic appendectomy (LA) is the most dominant method of current operative therapy and the gold standard technique in the elderly. Population based studies have shown a lower rate of complications and death in the elderly (2.4 vs. 0.5%) for open vs. LA in patients over the age of 65 years [8]. The high incidence of complicated appendicitis in elderly patients can lead to an increased rate of the open procedure. In the elderly, perforation can occur very quickly, supposedly because of degenerative blood vessel changes [9].

Adenocarcinoma of the appendix is the most common perforating cancer of the gastrointestinal tract. Anatomically, there are several reasons for this, namely the extremely thin subserosal layer, gentle submucosal and thin muscular layer of the appendix. Interestingly, in a large number of patients perforation does not have a significant impact on the outcome [4]. Generally, the clinical manifestations of appendicular tumors are similar to those of acute appendicitis, or a tumor may present as a palpable mass in the abdomen [4]. Both our patients presented with a perforation of the appendix with a classical clinical manifestation of acute abdomen, leading to wrong intraoperative approach. No recurrence in these patients shows that perforation of AA does not imply a worse prognosis.

Malignancies of the appendix are rarely diagnosed preoperatively or intraoperatively. Most tumors are identified only after histopathological examination of the resected specimen following appendectomy [1]. Advances in imaging techniques in the past 20 years have changed the way suspected acute appendicitis is evaluated. The presentation of acute appendicitis may be atypical, complicating the diagnosis. Plain abdominal radiographs are not routinely recommended for the evaluation of suspected acute appendicitis, or AA. Advances in ultrasound technology and the graded compression technique have improved the ability to visualize the appendix. The graded compression technique involves applying steady, gradual pressure to the right lower quadrant in an effort to collapse the normal bowel and eliminate normal bowel gas to visualize the appendix. In case of acute appendicitis, the appendix wall is immobile, non-compressible, and thickened with a diameter greater than 6 to 7 mm [10]. In elderly patients presenting with an appendicular lump, the differential diagnosis of malignancy should be kept in mind [9]. The urgency of the operation in our patients restricted the range of diagnostic procedures. Preoperative abdominal computerized tomography is mandatory in the elderly patients to provide an early diagnosis and to decrease unnecessary surgical exploration if acute appendicitis is suspected [11]. There are some controversies regarding the surgical therapy of adenocarcinoma of the appendix. Murphy et al. suggested that appendectomy was appropriate for accidentally intraoperatively identified tumors if the tumor was confined to the appendix, less than 2 cm in diameter, without obvious infiltration of the mesoappendix and the base of the appendix [12]. Arellano et al. conducted a retrospective study including 53,019 appendectomies; there were 44 (0.01%) cases with primary adenocarcinoma of the appendix, and a 12-year survival in 59% of these patients. Right hemicolectomy was suggested as elective treatment in this study. These authors concluded that preoperative diagnosis of appendiceal malignancies was very rare because of similarities of clinical presentations with appendicitis [13].

Conclusion

Our results suggest that in patients with appendiceal adenocarcinoma and an advanced stage of tumor, right hemicolectomy is a better choice than appendectomy alone. In elderly patients with clinical symptoms of complicated acute appendicitis, appendectomy should be done with intraoperative histopathological frozen section examination. Preoperative diagnosis of appendiceal adenocarcinoma is a challenge due to overlapping of symptoms with complicated acute appendicitis. Ultrasound is not a method of choice in the diagnosis in the elderly patients.
References


