Primary adenocarcinoma of the vermiform appendix is a rare clinical condition. It usually presents as an acute abdominal process or as a mass in the right lower quadrant. The gross pathologic and microscopic characteristics of this disease are the same as for colon carcinoma and carry a serious prognosis. We report a case of a 64-year old male patient presenting symptoms of acute appendicitis. The patient underwent laparoscopic appendectomy that revealed a severely inflamed appendix. Histopathological examination has shown that it was a poorly differentiated, high grade adenocarcinoma invading the periappendicular fat. According to the histopathological findings, a right hemicolectomy was further advised and performed. Right hemicolectomy is the generally recommended method of treatment for invasive adenocarcinoma of the appendix.

Key words: adenocarcinoma, appendix, laparoscopic appendectomy, right hemicolectomy

CASE REPORT

A 64-year old man reported to the Emergency Department with a sudden onset of acute abdominal pain, localized in the right lower quadrant, accompanied by nausea and vomiting. On examination, the patient was found to be feverish (body temperature 38.2°C), pulse was 60/min and blood pressure 105/55 mm Hg. Abdominal examination has shown guarding and tenderness in the right lower quadrant with rebound tenderness. Digital rectal examination revealed enlarged prostate but no masses were felt. Baseline blood tests were normal apart from an elevated white blood count (WBC 16,500/mm³), and C-reactive protein (CRP 67 mg/dL). The patient’s significant
medical history was tonsillectomy and comorbidities including type 2 diabetes mellitus, arterial hypertension and ischemic heart disease. The ultrasound scan revealed hypertrophy of the prostate gland. All other abdominal organs were found to be within normal limits and no free fluid within the peritoneal cavity was visualized on ultrasound. The patient was taken to the operating room for laparoscopic exploration which proved the clinical suspicion of acute appendicitis. Macroscopically, the appendix was swollen and phlegmonous with a little collection of cloudy peritoneal fluid that was present in the right lower abdomen (fig. 1). Laparoscopic appendectomy was performed using the ligasure system (Liga Sure Atlas, Covidien) for the division of the mesoappendix, along with endostapler closure (Echelon Flex Stapler, Ethicon Endo-Surgery) of the appendiceal stump. The peritoneal cavity was lavaged with saline and a drain was placed in the rectovesical pouch.

Pathological examination of the surgical specimen revealed a primary appendiceal adenocarcinoma (high-grade, partially signet ring carcinoma) diffusely infiltrating the appendiceal wall, extending to its mesentery and the surrounding adipose tissue (fig. 2). The neoplastic infiltrate was near the resection margin. Post-operatively, the patient did well and his drain was removed. Postoperative wound suppuration was observed and appropriate topical treatment was applied. He was discharged on the sixth postoperative day to return later for follow-up examinations.

Staging examinations including colonoscopy, chest X-ray and abdominal ultrasound were normal. According to the histopathological findings and the principles of oncologic surgery, the

Fig. 1. Gross picture of the surgical specimen

Fig. 2. Histopathologic examination slides of the surgical specimen

A – tubular adenocarcinoma admixed with inflammatory infiltrate. HE staining; magnification 40x; B – signet ring carcinoma infiltrating the appendiceal wall. HE staining; magnification 40x; C – positive staining for mucin within signet ring cells. Mucino-carmine staining; magnification 40x; D – immunopositivity for cytokeratin 20 (CK20) within signet ring cells; magnification 20x
patient subsequently underwent a right hemicolectomy. No residual carcinoma with negative lymph nodes was found (0/7 nodes).

The patient was discharged from the hospital 10 days later and referred for adjuvant chemotherapy. He received an oral capecitabine with good outcome. No long-term postoperative complications occurred.

**DISCUSSION**

Primary adenocarcinoma of the vermiform appendix is an uncommon neoplasm of the gastrointestinal tract that is found in approximately 0.5% of appendectomy specimens, and generally affects adults (6). Moreover, a primary signet ring cell carcinoma of the appendix is an exceedingly rare condition comprising only 4% of all appendiceal neoplasms (7). It usually presents as an acute abdominal process or as a mass in the right lower quadrant causing luminal obstruction of the appendix. Diagnosis of an appendiceal adenocarcinoma usually occurs postoperatively, after histologic examination of the surgically removed inflamed specimen. Preoperative detection of appendiceal adenocarcinoma is rarely feasible via direct visualization during colonscopic examination (8).

The International Classification of Diseases for Oncology (ICD-O) divides the tumors of the appendix into five categories: colonic type adenocarcinoma, mucinous adenocarcinoma, signet ring cell carcinoma, goblet cell carcinoma, and malignant carcinoid/adenocarcinoid (9, 10). Appendiceal carcinomas have a poorer prognosis than carcinoids. Signet ring cell carcinoma is considered to be an aggressive type of cancer. In 93% of cases, metastases to adjacent organs, lymph nodes, or to the peritoneal cavity are often present at the time of diagnosis (9).

The treatment of appendiceal carcinoma is controversial. Some authors report that appendectomy is a curative procedure for lesions confined to the mucosa and recommend right hemicolectomy only for advanced stages of appendiceal carcinomas (11, 12). Thus secondary right hemicolectomy with lymph node dissection should be considered in patients presenting with an undifferentiated adenocarcinoma, in those with lymphatic involvement and with significant invasion of the submucosa or threatened resection margin (13).

Generally right hemicolectomy is considered as the treatment of choice for all patients with non-metastatic adenocarcinoma of the appendix with invasion beyond the mucosa. Appendicectomy alone seems to be an ideal treatment only for in situ and localized cases. This thesis was supported by Nitecki et al. (14) who demonstrated a survival advantage of 29% for right hemicolectomy over appendicectomy alone in 16-year series of 94 appendiceal adenocarcinomas. This is compatible with the results from the multi-institutional Surveillance, Epidemiology, End Results (SEER) database, which reported worse survival after appendectomy alone for appendicinal lesions (both carcinoid and adenocarcinomas) > 2 cm in diameter (4, 7).

The treatment options for metastatic disease include systemic chemotherapy alone, hypothermic intraoperative intraperitoneal chemotherapy, cytoreductive surgery with peritonectomy, and a combination of treatments (15). Based on the similarities between this type of appendix cancer and colon cancer, the treatment with systemic chemotherapy, that is commonly used to cure colon cancer, is also often used for the „colonic-type” appendiceal cancer. Some of the most commonly used intravenous chemotherapy agents are 5-fluorouracil (5-FU), leucovorin, oxaliplatin (FOLFOX-4) (15, 16). In the future, novel targeted therapies, including the epidermal growth factor receptor (EGFR) inhibitors and anti-angiogenic agents in combination with systemic chemotherapy and regional treatment should be concerned. Adenocarcinoma of the appendix is rare and is often diagnosed in an advanced stage. Despite surgery and adjuvant treatment, the prognosis remains poor.

**REFERENCES**

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