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Prikaz slučaja

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MUCINOUS APPENDICEAL ADENOCARCINOMA AS A RARE CAUSE OF ILEUS — A CASE REPORT

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Summary: Although appendiceal tumours are rare, they may be encountered unexpectedly in any emergency abdominal operation. Many of these tumours are not recognized intraoperatively and are diagnosed only during formal histopathological analysis of an appendectomy specimen. We present a rare case of appendiceal adenocarcinoma which caused acute bowel obstruction. The patient presented with abdominal pain, distension and constipation. Abdominal radiography showed large bowel obstruction. On laparotomy the appendix was abnormal, with a tumor growing in to the ileal loop and creating a bowel obstruction. An appendectomy was performed with en_bloc bowel segment resection and end ileostomy. Pathology specimens showed that the primary neoplasm was the appendiceal mucinous adenocarcinoma infiltrating the ileal loop. Patient underwent a right hemicolectomy, and recovered afterwards with no evidence of local recurrence or metastatic disease as confirmed by the standard follow-up.

Physicians should keep in their minds that appendiceal neoplasm may have various and nonspecific presentations, but only an accurate diagnosis will lead to correct patients treatment, healing and long-term survival.

Key words: appendicular neoplasm, mucinous adenocarcinoma, bowel obstruction.

INTRODUCTION

Appendiceal carcinomas represent a relatively uncommon clinical entity making 0,4–1% of all gastrointestinal malignancies (1). Neoplasms of the appendix are found in 1% of all appendectomy specimens with a frequency of appendiceal adenocarcinoma in 0,1% of the specimen investigated.

According to the data from statistics registry in the United States collected from 1973 to 2004, **adenocarcinoma** was the most frequently diagnosed appendiceal neoplasm reported in 65% of cases (2). They are more common in men, with the highest incidence in the fifth decade of life (3). Appendiceal carcinomas are infrequently thought of preoperatively due to non-specific symptoms and are also rarely detected intraoperatively during any emergency or elective abdominal operation. Diagnosis of mucinous adenocarcinoma of the appendix usually occurs after histopathological examination of an appendicectomy specimen; about 0.7–1.7% of appendectomy specimens contain an appendiceal neoplasm (1). The right hemicolectomy is considered to be the treatment of choice for all lesions with invasion beyond the mucosa and is considered as recommended treatment for all patients with nonmetastatic adenocarcinoma of the appendix. The approximate overall 5-year survival rate was recorded in 55-60% of the patients with nonmetastatic disease after right hemicolectomy and varies with stage and grade (4, 5).

CASE REPORT

An 77 year-old male patient was admitted to Clinic for Emergency Surgery, Clinical Center of Serbia, with abdominal pain, vomiting and constipation. His past medical history included chronic intestinal obstruction episodes during the last few months. On admission he was thin, with normal body temperature, systolic blood pressure of 106 mm Hg, and pulse of

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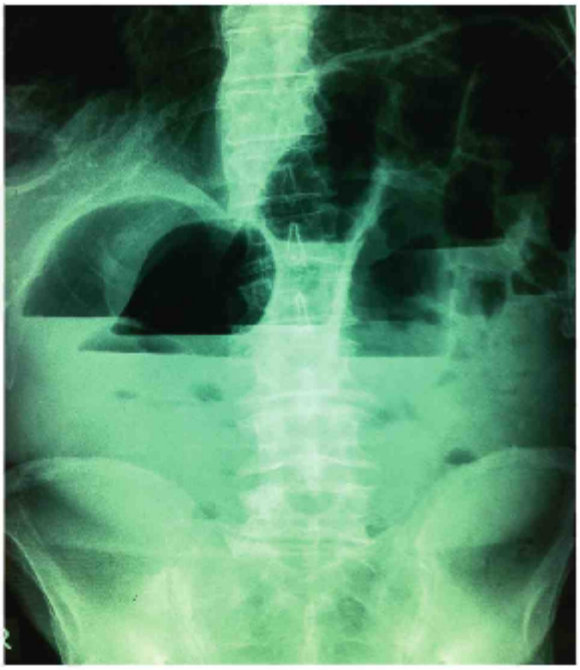

Figure 1. Emergency done abdominal radiography (XR) on admission showed dilated small bowel loops

100 beats per minute. On examination, the surgeon noted abdominal distension with tenderness. The White Cell Count was 13,4 x 10^9/l, Hemoglobin 101 g/l, Platelets 233 x 10^9/l, and blood biochemistry analyses were normal. Abdominal radiography (Figure 1) and Ultrasound (US) showed dilated small bowel loops. Intestinal obstruction dominated the clinical picture and these findings reached the diagnosis of acute bowel obstruction.

After a short period of resuscitation, a laparotomy was performed. The small bowel obstruction was found to be secondary to a tumor of distal ileum which grew in to the ileum from the top of the appendix. The appendix was noted to be abnormal but with no palpable ileocaecal lymph nodes. No distant lesions were seen. An appendectomy with en-block resection of distal ileum (200 mm in lenght) was performed and operation completed with end ileostomy. Postoperatively the patient made an uneventful recovery and he was discharged 7 days later.

The specimen was submitted to histopathological examination.

Macroscopic examination of the specimen showed the segment of small bowel length of 150 mm and appendix of 50 mm in length, which ingrew to the wall of the small intestine.

The cross-sections of the specimen showed vegetative tumor that completely filled lumen of the appendix, and established vegetations on the small bowel mucosa 70 x 75 x 80 mm in diameter, with the involvement of the 3/4 hose volume. The tumor was soft, honeycombed, of glossy gray-white appearance, about 20 mm away from the line of resection of the appendix.

Microscopic examination showed mucinous adenocarcinoma of appendix with infiltration of the small bowel. Metastatic tumour was not seen in 8 ileocaecal lymph nodes. The mucinous appendiceal adenocarcinoma was classified as stage A on Duke's staging system and as a TNM III (T4b, No, Mo) aaccording to TNM classification. Additionally, there was no evidence of malignancy in the appendiceal stump and no bowel margin resection. The patient was planned for a subsequent hemicolectomy.

One month after first operation, the patient underwent right hemicolectomy, with resection of 42 lymph nodes. On histopathological examination of the specimen, no malignancy was indentified, while the lymph nodes presented with only reactive inflammation. No evidence of local recurrence or metastatic disease was found by the standard follow-up, including tumour markers (CEA, CA-125, CA-19-9), and follow-up CT scanning of the abdomen (after 6 months and after 12 months). The patient was well for 18 months after the surgical treatment.

DISCUSSION

Mucinous adenocarcinoma is rare gastrointestinal malignancy.

Appendiceal adenocarcinoma is one histological type of the primary appendiceal neoplasm making 0,4 to 1% (3) of all gastrointestinal neoplasms (Table 1) (6, 7).

1. Mucinous adenocarcinoma
A. Mucinous cystadenocarcinoma
i. Invasive
ii. Non-invasive
B. Mucinous adenocarcinoma (non-cystic)
2. Intestinal-type adenocarcinoma
3. Signet-ring cell carcinoma
4. Neuroendocrine carcinoma
A. Small-cell neuroendocrine carcinoma
B. Large-cell neuroendocrine carcinoma
5. Mixed forms (including tumours with component of goblet cell carcinoid)
6. Undifferentiated carcinoma

Table 1. A modified classification of appendiceal carcinomas according to Misdraji and Young (7)

In a group of 1095 patients with primary appendiceal adenocarcinoma McCusker et al. found that from 56,0% of tumors classified as mucinous appendiceal carcinoma, 37,6% belonged to colonic and 6,4 % to signet ring cell subtype (8).

An unexpected clinical presentation of appendiceal neoplasm should be considered by clinicians. Appendiceal carcinomas are infrequently suspected preoperatively, appendiceal tumours present as acute appendicitis in 49% of cases, 5.4% as pelvic abscesses, 6.4% with gastrointestinal symptoms, 6.4% with bowel obstruction, 1,4% with carcinoid syndrome and rarely as RIF (Right Iliac Fossa) mass and inflammatory bowel disease (9).

Appendiceal neoplasms could be found incidentally at any emergency or elective abdominal surgery, and management plan should then be based on the intraoperative findings.

Early diagnosis and surgical intervention is very important in patients with bowel strangulation. Whatever the cause of intestinal obstruction was, we recommend using a midline vertical incision that provides the best access into the abdominal cavity. As part of the exploratory laparotomy, in addition to the routine thorough examination of the abdominal viscera, looking for deposits and lymphadenopathy, we suggest also careful inspection and palpation of the appendix and mesoappendix.

Murphy et al. suggested that if the tumour found incidentally at operation, was confined to the appendix, smaller than 2 cm, without evidence of mesoappendiceal involvement and not involving the base of the appendix, appendectomy is appropriate (1). Any neoplasm greater than 2 cm and involving the base of the appendix or mesoappendix should be considered for immediate right hemicolectomy in order to achieve the optimal outcome.

To be more detailed, right hemicolectomy is considered to be the treatment of choice for all lesions with invasion beyond the mucosa, and, appendectomy alone seems to be the ideal treatment for in situ and localized cases (10). For patients who were found to have appendiceal neoplasm on histopathological examination, if the lesion was benign or carcinoid less than 2 cm and confined to the appendix, then appendectomy alone is sufficient. Those who present with a perforation, though macroscopically removed, epithelial tumour should have baseline tumour markers (CEA, CA-125, CA-19-9), CT scan and colonoscopy (11).

The current and recommended surgical/oncological management for patients diagnosed with peritoneal metastasises or peritoneal carcinomatosis, consists of combination of cytoreductive surgery and intraoperative/intra-abdominal chemotherapy (12–17).

Primary mucinous adenocarcinoma of the appendix is often associated with synchronous and metachronous colorectal neoplasms (1). With this in mind, detailed examination of the abdomen during surgery is necessary.

Radiotherapy in general has no role in the treatment unless the resection margins were involved. Chemotherapy is offered to patients with positive lymph nodes (11).

The mucinous histology of appendiceal neoplasm is the best prognostic factor (18). McCusker et al. reported 5-year survival rates of 44% for tumors with the mucinous subtype (8). In a group of 94 patients Nitecki et al. found a survival of 100% for patients with tumors classified as Dukes A, 67% for Dukes B, 50% for Dukes C and 6% for Dukes D cancer (19).

CONCLUSION

The reported patient shows very rare entity of intestinal occlusion, due to mucinous adenocarcinoma of the appendix. In any case of acute intestinal obstruction, surgical exploration is necessary to remove the causes of obstruction. When diagnosis of appendiceal adenocarcinoma is confirmed, the treatment must be appropriate to the stage of the disease.

Sažetak

ILEUS IZAZVAN MUCINOZNIM ADENOKARCINOMOM APENDIKSA — PRIKAZ SLUČAJA

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Mada su tumori apendiksa retki, mogu se neočekivano sresti u toku bilo koje hitne hirurške intervencije. Mnogi od ovih tumora nisu prepoznati u toku operacije i bivaju dijagnostikovani tek patohistološkim pregledom preparata apendiksa. Prikazujemo redak slučaj adenokarcinoma apendiksa koji se klinički manifestovao kao akutna crevna opstrukcija. Pacijent se žalio na bol u trbuhu, nadutost i izostanak stolice. Tokom lapa-

rotomije nađen je tumorski izmenjen apendiks koji urasta u vijugu ileuma, stvarajući opstrukciju creva. Učinjena je apendektomija sa „enblok" segmentnom resekcijom tankog creva i terminalnom ileostomom. Pregled preparata na patologiji pokazao je da se radi o primarnom tumoru apendiksa, mucinoznom adenokarcinomu, sa infiltracijom vijuge ileuma. Izvedena je desna hemikolektomija i pacijent se oporavio, bez lokalnog recidiva i bez prisustva metastaza, što je potvrđeno na redovnim kontrolama.

Treba imati na umu da neoplazme apendiksa imaju različitu i nespecifičnu prezentaciju, a samo tačna dijagnoza vodi ka korektnom terapijskom pristupu, izlečenju pacijenta i dugoročnom preživljavanju.

Ključne reči: neoplazma apendiksa, mucinozni adenokarcinom, crevna opstrukcija.

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