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CASE REPORT

Mucous tumour of the appendix

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A – Research concept and design, B – Collection and/or assembly of data, C – Data analysis and interpretation, D – Writing the article, E – Critical revision of the article, F – Final approval of the article

Abstract

Background: In this paper we present a case of perforated mucinous neoplasm of the appendix. Diagnosis and treatment of this rare condition are briefly discussed.
Material and methods: A 42-year-old patient admitted to the General Surgery Clinic for elective surgery of left-sided inguinal hernia. In the postoperative period, severe pain in the lower abdomen persisted – qualified for laparotomy, during which an appendix tumor and a large amount of gelatinous bloody contents in the right lower abdomen were found. The appendix was removed and the peritoneal cavity was cleaned in the content. Histopathological examination revealed low grade appendiceal mucinous neoplasm (LAMN) pT4a NX M1b. No angiovasia or nerve trunk infiltration were found. At the top of the appendix a perforation with mucus around was noted. Postoperative course was uncomplicated. The patient was referred to a peritoneal carcinomatosis center where a right hemicolectomy with HIPEC therapy was performed.
Results: Mucous neoplasms of the appendix of low grade (LAMN) are rare tumors.
Conclusions: Mucous neoplasms of the appendix of low grade (LAMN) are rare tumors. The treatment of choice is surgical excision. The most serious complication of LAMN is a rupture with the escape of mucus into the peritoneal cavity.
Keywords: myxoma, pseudomyxoma peritonei, appendix, tumours

Introduction

Appendicitis tumours are rare. They are present in approximately 0.2%–0.3% of cases of appendicitis and 8%–10% of appendix tumours [1,2]. Low-grade appendix mucous tumour (LAMN) is a rare tumour that causes bulbous appendix enlargement due to the secretion of a large amount of gelatinous mucus [3]. This tumour is a rare occurrence found in 0.3% of appendicitis sections [4,5]. The average age at diagnosis of the disease is 70 years and it is more common in women [6]. The symptoms of LAMN are often atypical, discovered accidentally during other tests [7]. An extremely

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rare complication of LAMN may be the formation of pseudoperitoneal myxoma, a malignant tumour that is difficult to treat and fills the abdominal cavity with gelatinous contents [8].

Material and Methods

A 42-year-old patient was admitted to the General Surgery Clinic for elective surgery on a left-sided inguinal hernia. In the postoperative period, severe pain in the lower abdomen persisted, so he was qualified for a laparotomy, during which an appendix tumour and a large amount of bloody gelatinous contents in the right lower abdomen were found. The appendix was removed and the peritoneal cavity was cleaned of the contents. A histopathological examination revealed a low-grade appendiceal mucinous neoplasm (LAMN) pT4a NX M1b. No angiovasia or nerve trunk infiltration was found. At the top of the appendix a perforation with surrounding mucus was noted. The decision to perform a laparotomy was based on clinical signs of acute abdomen. No CT or ultrasound was performed before surgery due to an open laparotomy procedure with resection except for the healthy margin. The postoperative period was good. He did not report abdominal pain and other gastrointestinal disorders. On inspection visits his condition was good. The abdomen was soft and painless. The rhythm of bowel movements was within the normal range. The patient followed the surgical recommendations. The final effect of the surgical treatment was positive. The patient was referred to a peritoneal carcinomatosis centre where a right hemicolectomy with HIPEC therapy was performed. No peritoneal implants were noted.

Discussion

Appendicitis tumours are rare. They occur most often in the form of myxoma. They can arise as a simple cyst that forms while the lumen of the appendix is obstructed, accompanied by normal mucus production – gallstone, faecal stone, endometriosis, torsion of the appendix, intestinal tumour and even postoperative adhesion [9,10]. In the case of excessive mucous hyperplasia without the features of atypia, the lesions do not usually exceed 2 cm in diameter and rarely threaten to rupture. Mucinous cystadenoma (benign hyperplastic lesion) and adenocarcinoma mucosacus (malignant tumour) with features of advanced or moderate dysplasia may be symptomatic and perforated. They usually reach a size of about 6 cm to 25 cm [9,11–13]. They can be misdiagnosed as retroperitoneal tumours or right adnexa [7,14]. In most cases, they are detected accidentally, during other examinations within the abdominal cavity. Ultrasound and computed tomography can clearly show the size of the appendix tumour, inspection, the amount of mucus in

the lumen and anatomical relationships [15]. Computed tomography is more sensitive than ultrasound in evaluating tumours and detecting parietal calcifications, suggesting a diagnosis but only revealed in 50% of cases [16]. Surgical resection of low-grade appendicitis mucosal neoplasms is recommended [17]. The most important point in dealing with LAMN is to avoid rupture. Mucin, which fills the abdominal cavity, can lead to pseudomyxoma peritonei, a rare malignant tumour, characterized by the accumulation of a large amount of mucus in the peritoneal cavity. Although low-grade appendix mucosa is a slow-growing tumour, both LAMN and mucosal adenocarcinoma may progress to peritoneal myxoma [6,7]. In addition to the increase in CRP levels, elevated levels of markers (CEA, CA19-9) can be observed, suggesting hyperplastic etiology or the presence of concomitant changes [12,18]. The five-year prognosis for patients with a malignant appendix tumour complicated by pseudomyxoma peritonei is only 53% [19]. For benign lesions, the five-year survival rate is 100% [12,20]. Treatment of pseudomyxoma peritonei is difficult and currently relies on the HIPEC technique combined with peritonectomy [21]. HIPEC (Hyperthermic Intraperitoneal Chemotherapy) provides hyperthermia induction and direct intra-abdominal administration of chemotherapy in addition to intracellular drug secretion. It is used in patients with cancer dissemination to the peritoneal cavities, e.g. gastric cancer, cancer of the appendix, cancer of the colon, mesothelioma [22].

Results

Low-grade mucous neoplasms of the appendix (LAMN) are rare tumours.

Conclusion

Low-grade mucous neoplasms of the appendix (LAMN) are rare tumours. The treatment of choice is surgical excision. The most serious complication of LAMN is a rupture with the escape of mucus into the peritoneal cavity.

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