

Case Report

Case of an Low Grade Appendical Mucinous Neoplasia (LAMNS) Presenting as a Right Ovarian Mass

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Introduction

Appendiceal carcinoma has an incidence of 0.12 cases per 1,000,000 people per year [1]. Of all intestinal tumors, malignant tumors of appendix are less than 0.5%. They are classified into three groups: carcinoid tumors, mucinous cystadenocarcinomas and adenocarcinomas, based on architectural and cytologic features [2]. Appendiceal carcinoma present with acute right abdominal pain, and can also present as a palpable abdominal mass, acute intestinal obstruction or ascites. Most of them are diagnosed unexpectedly from histopathological analysis after a simple appendectomy.

That is why it is important for gynecologists and general surgeons to consider appendiceal mucinous cystadenomas in their differential diagnosis for a pelvic mass, because early recognition can prevent iatrogenic rupture and subsequent spilling of potentially malignant mucin-producing cells in a peritoneal cavity. In our case report, we present a low grade mucinous appendical tumor, diagnosed during surgery in an operating room.

Case Presentation

We present a case of a 64 year old Macedonian female, with complaints of a left sided pelvic pain. She was hospitalized at a University Clinic of Obstetrics and Gynecology – Skopje, a tertiary care teaching university hospital. After the admission, a laboratory and clinical researches were done. Tumor markers were in a different values, and also the full blood test. One year ago she was treated laparoscopic with right sided adnexectomy and the histopathology finding presented Fibroma of the right ovarium. Also she has a membranoproliferative glomerulonephritis since 2001, treated with therapy. The patient is in senium, with finished reproduction, two spontaneous births and a menopause 19 years ago. An adnexal mass was revealed on ultrasound and MRI, such as clearly demarked change in the region of right tuba 70x80 mm. She has an urge incontinently stage II, treated with medications. This patient was presented Infront of advisory board after the morning meeting. This case was presented at an expert consilium of the University Clinic for Obstetrics and Gynecology, which decided for a planned abdominal hysterectomy with left side adnexectomy for a treatment. During the operation, she was unexpectedly found to have an abnormally large

appendix with changed bright coloration, dilated and adhered to the peritoneum along with extensive adhesions between right tuba and pericolic adipose tissue of the cecum. General surgeon was consulted intra operatively to perform an appendectomy. The final histopathological report showed Low grade Appendical Mucinous Neoplasia (LAMNs), with low grade of malignancy.

Discussion

Mucinous cystadenoma of the appendix is an uncommon clinical finding. It is generally termed “mucocele” of the appendix, which simply refers to a cystic mass filled with mucin in a dilated appendix. The 4th edition of World Health Organization (WHO) classification had officially introduced “Low-grade Appendiceal Mucinous Neoplasm” (LAMN) as the appropriate name [3].

Retrospective studies have shown that up to 25% of patients harboring large lesions remain asymptomatic until the lesions are discovered incidentally during surgery. The most common symptoms, are severe right lower quadrant pain with a palpable abdominal mass present in 50% of cases [4-7]. Other symptoms that can be also present are nausea, vomiting, weight loss, appendicitis, urethral compression, lower gastrointestinal bleeding or intestinal obstruction.

The prevalence is higher in women than in men (4:1), with average age of 54 years old at the time of diagnoses [8,9]. Appendiceal mucinous adenomas can be particularly problematic for women as these tumors are commonly misdiagnosed pre-operatively as right-sided adnexal masses due to commonalities in clinical presentation. S LAMN generally grow slowly, and tend to produce the clinical picture of low-grade pseudomyxoma peritonei [10-13]. They tend to involve the appendix in a circumferential fashion with atrophy of the underlying lymphoid tissue. LAMN can proliferate outside the appendix in a malignant way, producing pseudomyxoma peritonei and even distant metastasis [14,15].

The morbidity/mortality associated with LAMN stems from rupture and intraperitoneal spread of mucin-producing epithelium, which may cause pseudomyxoma peritonei. As a result, gentle tissue handling during operation cannot be overemphasized. LAMN is associated with colon and ovarian malignancy, so intraoperative exploration of the entire gastrointestinal tract and ovaries in females is a very important part of the treatment. All gross peritoneal implants should undergo a biopsy and be removed with grading of the degree of epithelial atypia for prognostic purposes [16,17].

Conclusion

Appendiceal carcinomas can create a diagnostic dilemma due to non-specific clinical findings and inadequate imaging studies. They are usually asymptomatic and are diagnosed incidentally during surgery. Mucinous neoplasms with acellular mucin in the appendiceal wall or surrounding the appendix are associated with an

excellent prognosis and these cases should be managed similarly to appendiceal adenomas. Early recognition and careful intra-operative precautions must be taken to maintain the integrity of the appendix to prevent iatrogenic rupture and the spread of potentially malignant mucin-producing cells throughout the peritoneal cavity. The follow-up practice must be performed, because this tumor can coexist with other neoplasms. So, other investigations such as CT, US or colonoscopy are also recommended to detect other colon and ovarian malignancies.

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