

INCIDENTAL FINDING OF LOW-GRADE APPENDICULAR MUCINOUS NEOPLASM IN PERFORATED APPENDICITIS: A CASE REPORT

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Abstract: We are presenting a case of a 36 years old male patient with clinical signs for acute appendicitis. Contrast-enhanced abdominal CT finding revealed a small periappendicular abscess and the patient was subjected for laparoscopic appendectomy for complicated appendicitis. Histologic analysis of the specimen revealed a Low-grade Appendicular Mucinous Neoplasm (LAMN) with dimension of 1 cm at the appendicular tip with histologic presentation of mucin-producing cells.

Keywords: LAMN, perforated appendicitis.

1. INTRODUCTION

Appendicular mucinous neoplasms (AMN) are rare mucin-producing tumors of the vermiform appendix subdivided as low-grade and high-grade neoplasms, based on their present cell atypia. By definition, a low-grade appendicular mucinous neoplasm (LAMN) is a mucin-producing appendicular neoplasia with low-grade cell atypia, but without a presentation of infiltrative invasion, contrary to the appendicular mucinous adenocarcinoma. Their clinical presentation is non-specific and are often discovered incidentally. However, they can cause complicated appendicitis even in up to 29% of the cases. In the United States only 1000-2000 cases are been reported per year. When clinical signs of acute appendicitis are absent, the diagnosis of LAMN can be challenging. Fortunately, the diagnostic imaging tools in most cases demonstrate abnormal presentation of the appendix and therefore an operative management (appendix removal) is recommended.

LAMN treatment varies from simple open or laparoscopic appendectomy alone, close and careful surveillance for patients with localized periappendicular disease following initial surgery up to extended cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC) for disseminated peritoneal disease.

2. CASE PRESENTATION

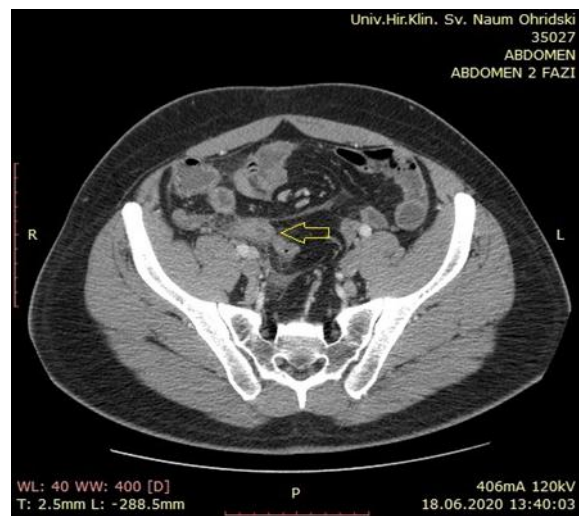
Thirty six years old male patient referred to our emergency abdominal department for clinical signs of acute appendicitis that last in the past 20 hours. The physical examination revealed abdominal tenderness over the ileocecal valve with positive sings for acute appendicitis. Laboratory test showed elevated Leukocyte count of $11.5 \times 10^9/l$ ($3.8 - 5.8 \times 10^9 /L$), elevated CRP level of 58.1 mg/l ($0 - 5.0 \text{ mg/L}$) and total serum bilirubin of 33.8 mmol/L ($3.4 - 20.5 \text{ mmol/L}$). The rest of the laboratory values were within the normal limits.

Plain abdominal x-ray (**Figure 1**) and abdominal ultrasound were indicated. The ultrasound did not show direct signs for acute appendicitis. Contrast-enhanced computer tomography of the abdomen was done. It revealed a periappendicular abscess with dimensions of the appendix around 4 cm (**Figure 2**).

Figure 1. Plain abdominal x-ray

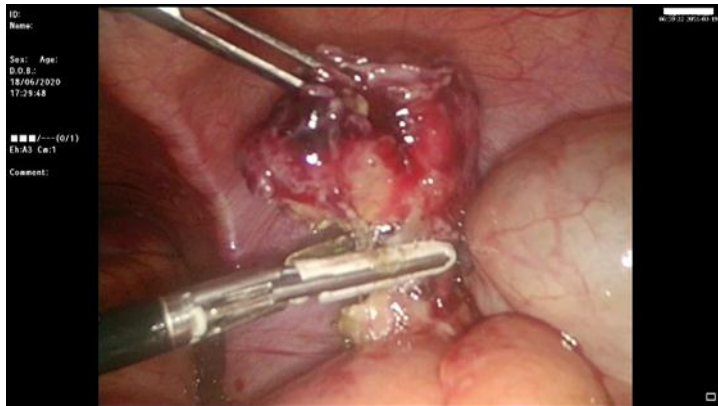


Figure 2. CT finding of small periappendicular abscess- yellow arrow



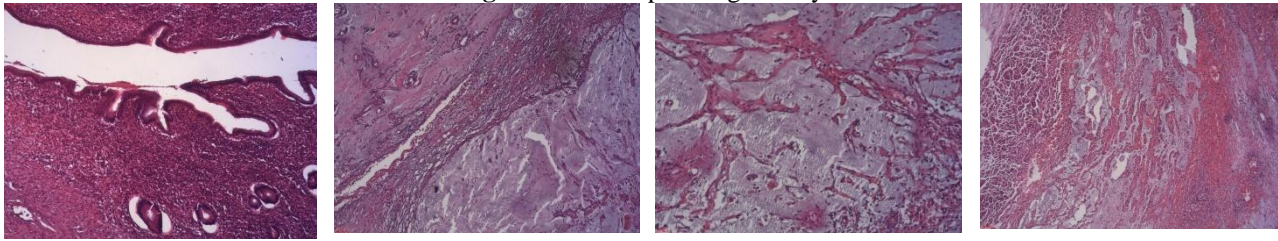
The patient was offered a standard three - port laparoscopic appendectomy due to the probable preoperative diagnosis of complicated appendicitis. During the procedure, a small periappendicular abscess was confirmed with a free perforation of the appendix (**Figure 3**). Appendectomy followed and the operation finished without incidents. The postoperative period was uneventful and the patient was released home on postoperative day 4. The macroscopic pathology specimen analysis revealed vermiform appendix with a length of 9.5 cm and dilated lumen with diameter of 1 cm. The tip of the appendix showed cystic changes with edematous wall.

Figure 3. Intraoperative finding



Microscopic examination presented a mucinous neoplasm at the appendicular tip with dimensions of 1 cm with low degree of malignancy. There were no tumor cells in the lymphovascular vessels and mucin production was seen intra and extracellularly. The final finding defined a low-grade appendicular mucinous neoplasm with pTNM = pTis, pNx (according to UICC, 8th ed.) (**Figure 4-7**). An oncologist was consulted for eventual further adjuvant treatment. According to the stage of the neoplasm, a regular long-term follow up was recommended.

Figure 4-7. Histopathologic analysis



3. DISCUSSION

From etiological aspect, an acute appendicitis can be caused by an appendicular tumor and even can cause appendicular perforation (1). Different clinical patterns can occur in advanced stage of the disease: appendicitis-like symptoms, chronic abdominal pain, weight loss, anemia or onset of abdominal wall hernia.

From pathological point, the diagnosis of LAMN depends primarily on the presence of mucin in the cells. Immunohistochemical staining characteristics for LAMN are: 100% positivity for CK20, usual negativity for CK7 in 71% of cases and positivity for MUC5AC (86%) and DPC4 (100%).

The treatment for AMN varies according to the disease stage, tumor size and its distant spread. Due to the low incidence of nodal spread (less than 2%) for well-differentiated AMN's (including LAMN), simple appendectomy is curative. In cases where the tumor is larger than 2 cm, invades the muscularis propria, is low-differentiated, involving the periappendicular area or with positive margin after appendectomy, a right hemicolectomy is indicated. More advanced stages of AMN with present peritoneal metastases are treated with debulking or cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). The combination of CRS and HIPEC is proved to prolong 5-year survival rate up to 86%.

In cases of LAMN with present peritoneal mucin spillage, the data for the type of surgery is controversial. Appendectomy alone or right hemicolectomy are not recommended due to high probability of future disease progression and peritoneum involvement and cytoreductive surgery for tumors with localized mucin presence is advised.

4. CONCLUSION

Our patient finished the first scheduled postoperative surveillance from the follow-up period with normal findings for serum tumor markers (CEA, Ca 19 - 9 and CA - 125) and the abdominal CT scan. The recommended plan for his follow-up due to his age and the nature of the disease is physical examination, tumor markers and CT scan controls every 3 months during the first year after the surgery, then every 6 months in the next 4 years of postoperative period and a yearly examinations afterwards.

The low incidence and data insufficiency for the standard treatment of LAMN should not force the clinicians to forget about it. In contrary, it should raise awareness about its presence, the incidental but serious diagnosis of itself in order to provide higher rate of preoperative diagnosis and, at the same time, to allow adequate surgical treatment in order to enable a positive outcome.

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