APPENDICEAL MUCINOUS NEOPLASMS: A CASE REPORT AND REVIEW OF LITERATURE

Jia Wu, MD
Xin Huang, MD
Wentun Yao, MD
Dongtao Liu, MD
Junwen Ma, MD
Liya Huang, PhD

Appendiceal mucinous neoplasms (AMNs) are uncommon tumors accounting for less than 1% of all cancers, which include a heterogeneous group of diseases with varying malignant potential. Pseudomyxoma peritonei (PMP) is a clinical syndrome characterized by progressive accumulation of mucinous tumor throughout the peritoneal cavity (Carr et al., 2017). Because of difficulty in recognition during diagnosis, it is important to identify special clinical manifestations in AMNs patients. We here report a case of a patient diagnosed with AMN.

Case Presentation
A 60-year-old man was referred to our hospital because of ascites of unknown origin for 4 years. During these 4 years, he had abdominal
distension and been diagnosed with liver cirrhosis. He had undergone relevant treatment, such as ascites drainage and diuretics, but without satisfactory effect. The patient had an otherwise insignificant previous medical history. On admission to our hospital, physical examination showed a fluid-filled abdomen; however, shifting dullness was not obvious. The abdomen was soft, with no direct tenderness or rebound tenderness.

The patient had stable vital signs. Blood and urine analyses were normal. Electrocardiography, chest radiography, and arterial blood gas analysis were also normal. Abdominal computed tomography (CT) suggested that there was a huge cystic lesion in the patient’s abdomen and it seemed to be a lump containing solid and cystic components arising from the appendix (Figure 1). The patient also underwent colonoscopy examination, which showed that the colon and rectum as well as orifice of vermiform appendix were normal (Figure 2).

To identify the cause of the illness, subsequent exploratory laparotomy was performed. During the operation, a large amount of yellow and jelly-like turbid liquid (about 4,000 ml) was observed in the abdominal cavity (Figure 3). After eliminating these abnormal substances, an ileocecal appendix lump (Figure 4) appeared. The appendix lump was removed subsequently.

Microscopically, the pathology of the appendix lump tissue (Figure 5) suggested hyperplasia of the mucous columnar epithelium and the mucus lake, which was formed by a large amount of abnormal mucus. The pathology of peritoneal nodules showed that there was a large amount of mucous tissue in the hyperplastic fibrous tissue. The final diagnosis was low-grade AMN (LAMN) accompanied with PMP.

After confirming the diagnosis, the patient underwent a cytoreductive surgery (CRS), and 1 month after operation, repeat CT scan showed no abnormalities (Figure 6). Three months after CRS, the patient
underwent hyperthermic intraperitoneal chemotherapy (HIPEC) with paclitaxel and cisplatin. Furthermore, we conducted telephone follow-up every 6 months until April of this year, and the patient was alive and in good condition.

**Discussion**

AMNs are uncommon neoplasms that are detected in only 1% of appendectomy specimens (Fournier et al., 2017). Because of the low-incidence rate and atypical clinical presentation, these patients may be misdiagnosed in clinical practice. Most cases are diagnosed incidentally at the time of appendicitis surgery or a screening CT. In this report, the patient was diagnosed as having ascites caused by cirrhosis for 4 years. However, the diagnosis of AMN was eventually confirmed by CT and surgery combined with pathology. Therefore, it is crucial to distinguish between ascites and pseudoascites (PA).

PA is an uncommon cause of abdominal distention mimicking ascites. When PA is caused by large abdominal cysts, it is difficult to distinguish from simple ascites because of the presence of an ultrathin wall and large liquid collection (Rossi et al., 2019). Shifting dullness and a ruler-pressing test are typical clinical signs of ascites upon physical examination. The typical radiological signs of PA—the fact that the loops appear conglomerated in the center of the abdomen, or the fact that there may be aspects of septations in the fluid collection—are not a consistent finding at imaging and also they are not specific (Camilon & Chilstrom, 2014; Nett, Vo, & Chapman, 2015; Shafi, Malla, & Reshi, 2009). Ascites from liver cirrhosis appears on the basis of the symptoms and signs of liver function damage. Although surgery combined with pathology examination is the criterion standard for diagnosis, it is beneficial for patients to undergo a differential procedure before an invasive surgery.

According to the new 2016 consensus statement from the Peritoneal Surface Oncology Group International (PSOGI), mucinous neoplasms are classified as follows (Carr et al., 2016): LAMN, high-grade AMN (HAMN), mucinous adenocarcinoma, and poorly differentiated mucinous adenocarcinoma with signet ring features. LAMN extends beyond the mucosa into the appendiceal wall but without infiltrative invasion and with low-grade cytologic atypia (Carr et al., 2016). Regardless of the degree of cellular atypia, both LAMN and HAMN can develop transmural extension or can rupture. When this occurs, the patient is at risk for developing PMP (Bartlett et al., 2019).

This patient was finally diagnosed with LAMN accompanied with PMP by pathology examination. In recent years, a consensus both domestically and internationally recommends the integrated therapy of CRS and HIPEC as the standard treatment for PMP (Li, Yu, & Liu, 2014; Yang et al., 2019). Therefore, this patient received HIPEC treatment 3 months after surgery, rather than immediately following CRS. In subsequent telephone follow-up every 6 months until April of this year, the patient did not experience any discomfort or recurrence. These results indicate that similar to the effect of immediate HIPEC after surgery, treatment with HIPEC 3 months after surgery may still achieve satisfactory results. Further large-scale, multicenter clinical studies are required to explore this point.

**Conclusion**

It is of great importance to identify ascites related to PA for patients diagnosed with AMN accompanied by PMP. Firmly mastering basic knowledge and skill of physical examination and radiographic imaging is a necessary quality for modern clinicians. In addition,
early and accurate diagnosis will provide a clear direction for early treatment, thereby laying a solid foundation for improving the patient’s prognosis.

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