



Primary mucinous borderline tumor of the vermiform appendix mimicking ovarian carcinoma; case report

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Summary

Introduction: Primary adenocarcinoma of the vermiform appendix is a very rare entity. Borderline mucinous tumor of the appendix is a much more rare disease. **Case:** We present a 71-year-old woman with primary mucinous borderline tumor of the vermiform appendix. She was misdiagnosed as an ovarian carcinoma patient and operated on by a gynecologic oncology team. Her frozen section analysis of an appendiceal mass resulted as borderline tumor of appendix vermiformis, and right hemicolectomy, as is advised in invasive colon tumors, was not carried out. **Conclusion:** Borderline mucinous tumor of the appendix vermiformis should be kept in mind in patients who have pseudomyxoma peritonei during surgery. Tumoral resection may be the definitive therapy in these patients.

Key words: Pseudomyxoma peritonei; Borderline; Tumor of appendix vermiformis.

Introduction

Primary adenocarcinoma of the vermiform appendix is a rare entity and usually discovered by the pathologist in appendectomy specimens. Neoplasms of the appendix are found in 1% of all appendectomy specimens [1]. Primary appendiceal carcinoma varies from 0.01 to 0.2 per 100,000 persons per year [2, 3]. Primary borderline tumor of the vermiform appendix has been reported in very few case reports and is a new entity. Treatment choices and prognostic factors are still debated for this type of neoplasm.

Case Report

A 71-year-old woman (G3/P3) was admitted to our hospital with the complaint of increasing abdominal girth. Ultrasonographic evaluation revealed a right pelvic complicated mass located in the ovarian region. Pelvic examination also revealed a right ovarian conglomerated mass; the uterus and left ovary were free and the Douglas pouch was filled with ascites. CA125, 19-9 and 15-3 were all in normal ranges preoperatively.

The patient was diagnosed as having ovarian carcinoma and staging surgery was planned. Our clinic's gynecologic oncology team prepared the patient for surgery and all of her preoperative tests were within normal ranges.

A laparotomy was performed and the pelvic cavity was found to be full of mucin, with a right pelvic mass consisting of the appendix vermiformis and right ovary. All abdominal organs were covered with mucin and debris. The mucin was cleared with repeated washings. Staging surgery consisting of total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and bilateral pelvic and paraaortic lymphadenectomy was carried out. General surgeons were invited to the

operation room to evaluate the appendix vermiformis and an appendectomy was carried out. The lesion was a cystic mass, with a mucinous part of 2 cm in diameter on the outer portion (Figure 1). Intraoperative pathologic evaluation of appendectomy material with frozen section was reported as "mucinous appendiceal tumor of unknown behavior" and the operation ended after this pathologic diagnosis. Pathologic evaluation of paraffin-embedded specimens (the entire lesion was submitted to microscopic evaluation) revealed a mucinous tumor with low-moderate degree atypia, and no stromal invasion or epithelial cells in the mucin pools outside the appendix (Figure 2); the case was diagnosed as "mucinous tumor of uncertain malignant potential (borderline mucinous tumor) of appendix vermiformis". There was no tumor or metastasis in the other ovary, uterus or lymph nodes. The right ovary was free of tumor but there were some tumoral adhesions on it arising from the appendix mucinous cyst. There was no tumor on the colon side (base) of the appendix vermiformis. A decision for follow-up without adjuvant therapy was decided by the tumor board, and the patient is planned to have a 3-month period of follow-up visits.

Discussion

Primary adenocarcinoma of the appendix is a rare neoplasm diagnosed in less than 0.5% of all gastrointestinal system malignancies [1]. Borderline mucinous tumor of the appendix is a much more rare and interesting entity. Treatment regimens and the type of surgery needed are still debated [4]. The extent of surgery may differ from simple appendectomy to right hemicolectomy with regional lymph node dissection. Prognosis and survival figures for this type of borderline tumor and the effect of surgery are still controversial [5, 6]. Gynecologic oncologists and also colon surgeons may misdiagnose these patients because of interfering symptoms and findings. Borderline mucinous tumor of the appendix vermiformis should be kept in mind in patients who have pseudomyx-

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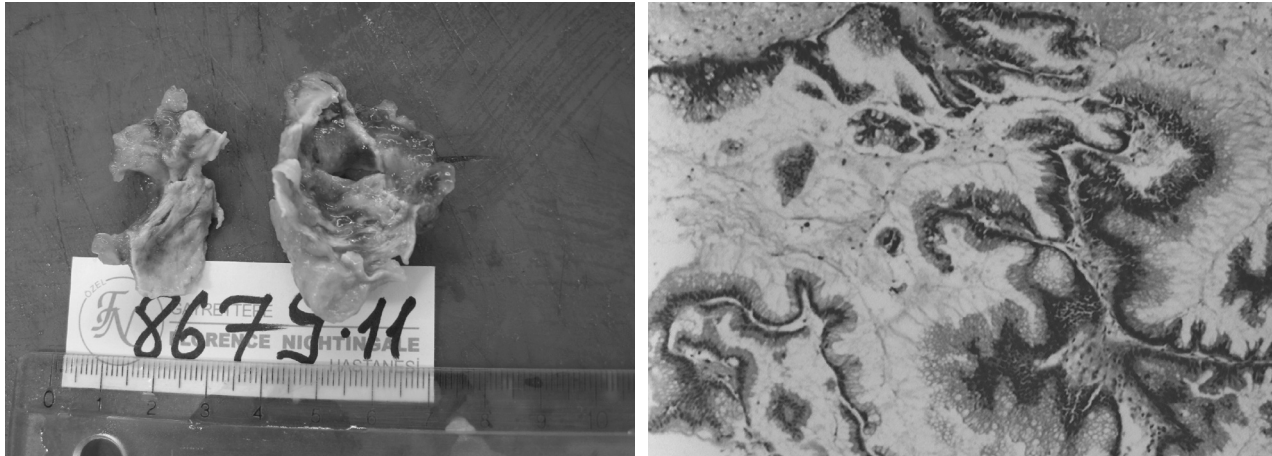


Figure 1. — Macroscopy of the specimen. Mucinous mass on the wall of the appendix.

Figure 2. — Pencil-shaped nuclei characteristics of adenomatous epithelium are evident, with low-moderate degree atypia (HE, x 100)

oma peritonei during surgery, as well as during intraoperative pathologic evaluation. The pathological diagnosis must be made with nearly all the specimen for microscopic evaluation. Tumoral resection may be the definitive therapy in these patients.

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