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Case Report

Rare Case of Appendiceal Signet-ring Cell Carcinoma Mimicking Advanced Ovarian Cancer

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Abstract

Primary signet-ring cell carcinoma of the appendix is extremely rare, and because the symptoms are vague and nonspecific, obtaining a preoperative diagnosis is difficult. It cannot be detected using imaging modalities because its radiologic features are undefined, and its appearance on both computed tomography and ultrasound images is similar to that of nontumorous appendicitis. Usually, a diagnosis is made after surgery for other diseases involving the removal of the appendix. The right hemicolectomy is suggested for invasive adenocarcinoma; however, diagnosing primary signet-ring cell carcinoma before surgery and managing tumors with adequate surgical resection remain challenging. We present a case of a large ovarian tumor with peritoneal carcinomatosis diagnosed with advanced ovarian cancer but subsequently revealed through pathology to be primary appendiceal signet-ring carcinoma. Detecting appendiceal signet-ring carcinoma warrants additional surgical management, and treating it involves chemotherapy. This rare disease should be kept in mind when encountering abdominal right lower quadrant pain and tumors to allow for a full treatment plan.

Keywords: Abdominal distension, adnexal tumor, appendix, ovarian cancer, signet-ring cell

Introduction

The incidence of primary carcinoma of the appendix is extremely low, affecting approximately 1–2/100,000 people each year in the United States.[1] The signet-ring cell subtype is particularly rare, comprising 4% of all appendiceal carcinomas.[2] Symptoms of appendiceal cancer are often nonspecific and most commonly present as right lower quadrant abdominal pain; it can mimic several diseases originating from the pelvic cavity, such as acute appendicitis, ovarian cancer, or other diseases related to the adnexa. Hence, primary cancers of the appendix are frequently diagnosed after surgery for appendicitis, presumed ovarian malignancies, or other reasons.[3] In this article, we present a patient with an initial diagnosis of advanced right ovarian cancer. She underwent suboptimal debulking surgery because of extensive

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disseminated peritoneal metastasis. The final pathology revealed primary signet-ring cell carcinoma of the appendix.

Case Report

A 68-year-old multiparous woman with no known medical or surgical history complained of abdominal distension for 1 month. She denied a loss of body weight, easy satiety, vomiting, abdominal pain, constipation, bloody stool, pencil stool, or irregular vaginal bleeding. On pelvic examination, a mass greater than the size of a fetal head was palpated over the right lower quadrant of her abdomen. Ultrasound displayed a cystic mass arising from the right pelvic cavity with a solid component measuring 15.8 cm × 10 cm [Figure 1]. Due to the impression of ovarian cancer, computed tomography (CT) was performed, and blood was drawn for tumor markers. The CT scan showed peritoneal seeding and a right ovarian tumor densely adhered to the uterus, bowel, and omentum [Figures 2 and 3]. No gastrointestinal tract tumor was suspected and no lymphadenopathy was detected. Elevated tumor markers, namely carcinoembryonic antigen (CEA; 750.3 ng/mL), cancer antigen 125 (CA125; 841.6 U/mL), and carbohydrate antigen 19-9 (CA19-9; 7328.3 U/mL), were noted. Since no gastrointestinal tract lesion was identified by CT scan, panendoscopy and colonoscopy were omitted. She underwent cytoreductive surgery for advanced ovarian cancer. During surgery, the adhesion between the appendix and ovarian tumor was inseparable, and an appendectomy was performed. The contralateral ovary was also involved. A frozen section of the ovarian tumor [Figure 4]was identified as a signet-ring cell tumor. Malignancy originating from the gastrointestinal tract was impressed. Because of extensive peritoneal carcinomatosis, suboptimal debulking surgery was conducted. Subsequent panendoscopy and colonoscopy both resulted in negative findings. The final pathology report revealed appendiceal adenocarcinoma of a signet-ring type [Figure 5] with bilateral adnexa, uterus, and omentum metastasis. The appendiceal tumor was immunopositive for CK7 [Figure 6] and CDX-2 [Figure 7].

After surgery, she received six courses of chemotherapy with irinotecan hydrochloride trihydrate, folinic acid, and fluorouracil. When her CEA level had decreased to 17.8 ng/mL, she rejected further treatment. Eight months after the last course of chemotherapy, she presented to the emergency room (ER) with dyspnea, abdominal distension, and bilateral leg edema. A thorough examination revealed massive ascites with spontaneous bacterial peritonitis and acute renal failure. She opted for palliative treatment and died of septic shock 10 days after her admission to the ER.

Discussion

The incidence of signet-ring cell carcinoma of the appendix is less than 5% of all appendiceal tumors, and the clinical symptoms are indistinguishable from those arising from the right lower quadrant abdominal cavity.

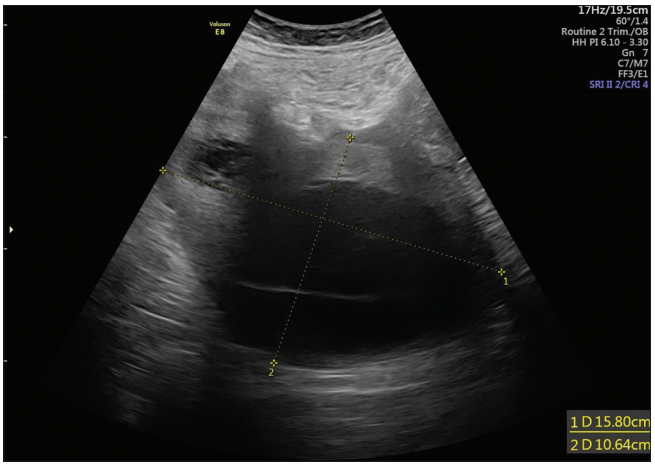

Figure 1: Transabdominal ultrasound showed a large, complex thick-walled cystic mass with solid components and septum arising from the right pelvic cavity

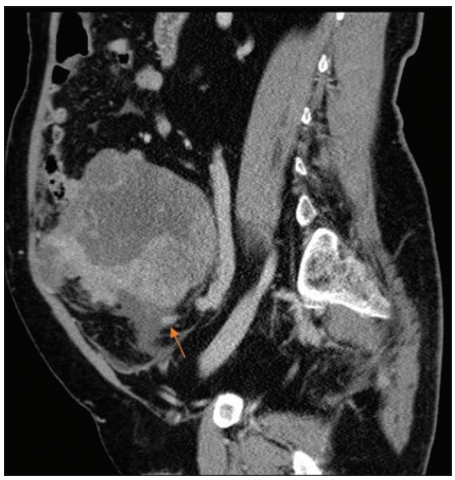

Figure 2: Sagittal contrast-enhanced computed tomography image showed a large cystic mass in the right abdomen, with solid and cystic components densely adhered to the uterus, omentum, and bowel loops. The arrow indicates a nondistended appendix with a thin wall just below the mass

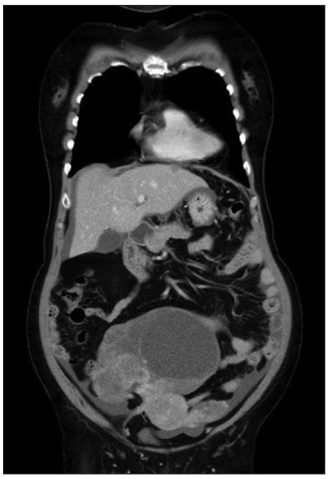

Figure 3: Axial contrast-enhanced computed tomography image showed that the cystic mass arose from the right adnexa of the uterus. The arrow indicates a nondistended appendix with a thin wall sitting below the mass

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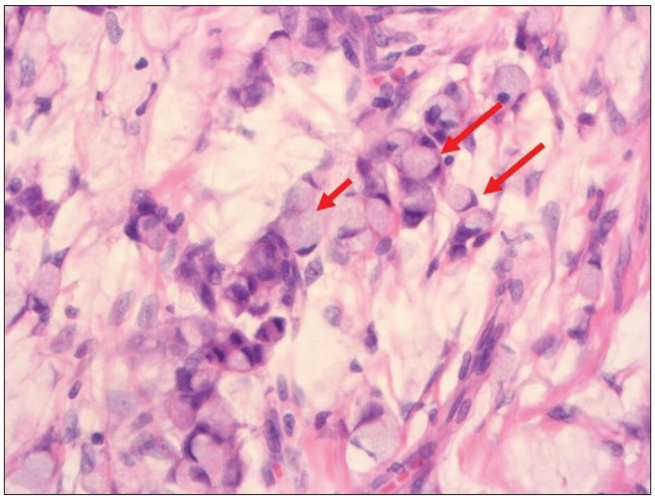

Figure 4: Ovarian tumor with tumor nests infiltrating in a fibromyxoid stroma. The arrows indicate signet-ring cells (H and E, ×400)

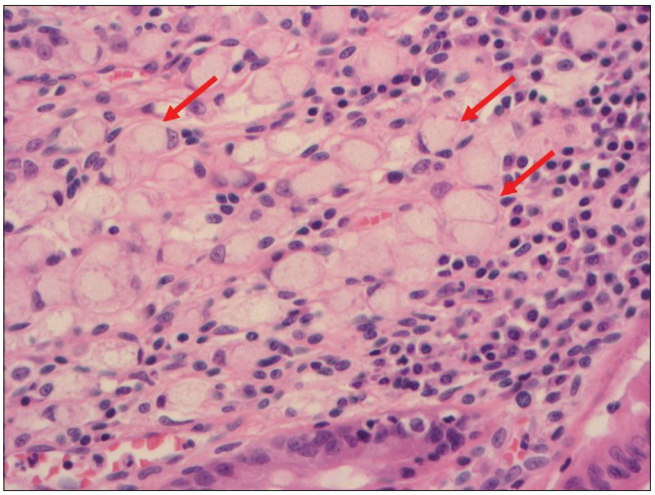

Figure 5: The arrows indicate signet-ring cells in the appendix (H and E, ×400)

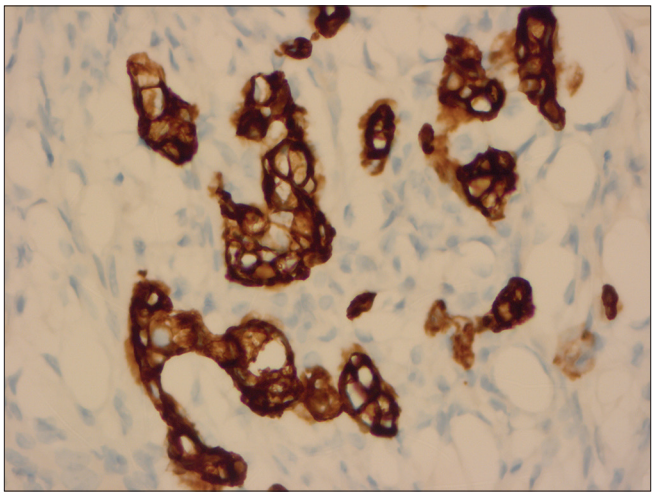

Figure 6: The tumor cells were reactive to CK7

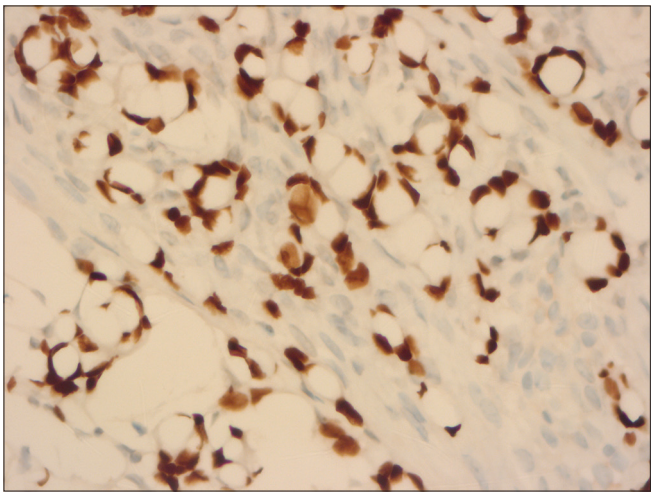

Figure 7: The tumor cell also revealed nuclear staining for CDX-2

The most common clinical complaint is abdominal pain in the lower right quadrant, which is similar to that of acute appendicitis.[4] Preoperative detection can be used to design an appropriate surgical resection, but making an accurate diagnosis is not often possible before surgery.[5,6] Radiologic techniques, such as CT of the appendix, are nonspecific for appendiceal cancer.[6] Cho proposed that patients with concentric hypoechoic mucosal thickening and certain clinical presentations, such as potential ovarian or peritoneal metastasis or submucosal infiltrative tumors, including signet-ring cell carcinoma, should be examined to obtain another diagnosis.[5] Appendiceal sonography may be done when large pelvic tumors arising from the right pelvic cavity are seen. If the ultrasound image of the appendix shows a concentrically thickened wall with a hypoechoic submucosal and muscle layer, then signet-ring cell carcinoma should be considered in the differential diagnosis. No specific CT findings distinguish signet-ring cell carcinoma from other histological types, except those that indicate a spread to adjacent organs (76%) at presentation.[7] An accurate diagnosis and precise staging warrant a right hemicolectomy, a suitable surgical treatment.[8] Because of its aggressive behavior, close follow-up is necessary after cytoreductive surgery and chemotherapy.

Our patient complained of abdominal fullness, which is nonspecific for appendiceal tumors. The correct diagnosis was determined only after debulking surgery, which was initially performed for ovarian malignancy. Although she received six courses of chemotherapy, the disease failed to respond. This clinical circumstance is consistent with the knowledge of signet-ring cell carcinoma, and a preoperative diagnosis remains a challenge for appropriate surgical planning.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information

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to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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