Mucinous Adenocarcinoma of the Urachal Remnant with Pseudomyxoma Peritonei

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INTRODUCTION

We report a rare case of urachal carcinoma causing pseudomyxoma peritonei (PMP) in a patient who presented with symptoms of chronic urinary retention. It is very unusual for urachal carcinoma to present in this way.

CASE REPORT

A 63-year-old man presented with suprapubic pain, terminal dribbling, a suprapubic mass, and microscopic hematuria. Flexible cystoscopy showed a mass in the dome of the bladder (Figure 1). Magnetic resonance imaging revealed a cystic mass in the dome of the bladder, but separated from the bladder with fluid, debris, and nodularity within the abdomen and the pelvis. The patient underwent laparotomy, where extensive PMP was discovered. Thereafter, he underwent partial cystectomy, omentectomy with excision of the tumor.

Histological examination revealed mucinous adenocarcinoma of the urachal remnant with metastases to the omentum (Figures 2 and 3). The patient underwent further debulking of the tumor with limited peritonectomy. At 18 months follow-up, recurrence occurred in the liver and the patient underwent further resection.

DISCUSSION

Urachal carcinoma is a rare neoplasm, accounting for 0.01% of all malignancies and 0.34% of the bladder carcinomas.[1,2] Urachal carcinoma is usually seen in male

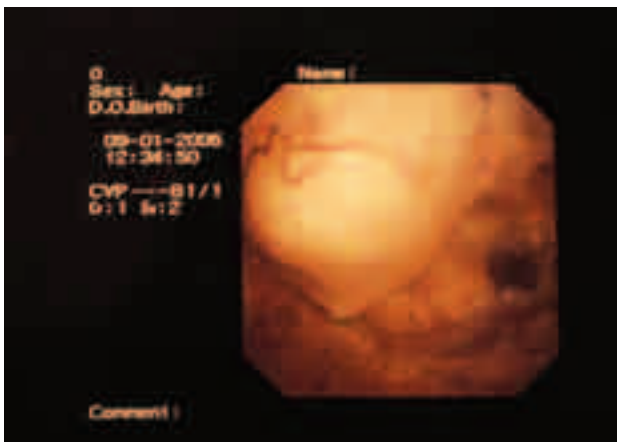

Figure 1. Cystoscopy showing tumor bulging into dome of the bladder.

Figure 2. Low power view of section of the bladder showing the interface between the normal bladder wall on the left and Urachal carcinoma on the right.

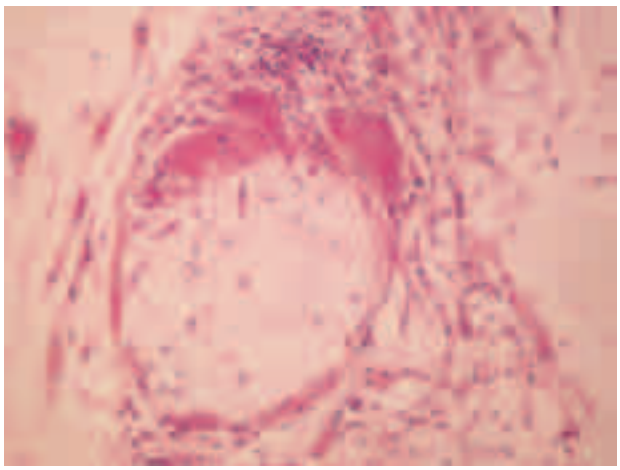

Figure 3. High power view of a slide showing nests of mucinous adenocarcinoma cells. Note the large quantities of extracellular mucin.

patients between 40 and 70 years of age.[3] The prognosis is poor; the tumor produces few symptoms until it is advanced. About 95% of patients have muscle invasion or metastatic cancer at the time of diagnosis.[4] Typically, the cancer spreads locally to the abdominal wall, and the peritoneum.[1,2] Although histological analysis reveals mucin production in about 75% of cases, mucinuria occurs in about 25% of patients.[5] The radiographic finding of supravesical calcification associated with a bladder lesion is pathognomonic for urachal adenocarcinoma.[3]

Pseudomyxoma peritonei is characterized by dissecting gelatinous ascites and peritoneal implants secreting copious amounts of extracellular mucin. Pseudomyxoma peritonei causes scalloping of the margins of the abdominal viscera, with septation of fluid collection with or without calcification. Progressive disease of the peritoneal cavity and compression of abdominal viscera are common, leading to wasting and repeated bowel obstruction.

Urachal carcinoma is almost uniformly resistant to chemotherapy or radiotherapy. Treatment for urachal carcinoma is surgical, usually en bloc radical cystoprostatectomy with wide excision of the urachus and umbilicus. Partial cystectomy with urachectomy can be as effective as more radical excision and causes less morbidity (5-year overall survival rate of 43%).[3,5]

This case highlights a rare presentation of urachal carcinoma with PMP in a patient with bladder outflow obstruction. It is a difficult tumor to treat and generally has a poor prognosis. Although there is little literature on the subject, some reports of less radical surgery suggest reduced morbidity with effective oncological results.

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CONFLICT OF INTEREST

None declared.

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