Cystic mucinous tumors of the urachus: carcinoma in situ or adenoma of unknown malignant potential?

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FAHED AC, NONAKA D, KANOFSKY JA, HUANG WC. Cystic mucinous tumors of the urachus: carcinoma in situ or adenoma of unknown malignant potential? The Canadian Journal of Urology. 2012;19(3):6310-6313.

Mucinous cystadenocarcinomas of the urachus are rare. Mucinous benign or premalignant tumors are even rarer, yet pose a challenge in diagnosis and management. We report a case of a 66-year-old man with lower abdominal pain who had a large cystic tumor at the dome of the bladder. En-bloc resection of the tumor with partial cystectomy revealed mucinous cystadenocarcinoma in situ. We reviewed the characteristics of all seven previously reported cases. These tumors are pre-malignant and can cause significant morbidity and mortality. They need to be treated similar to conventional mucinous cystadenocarcinoma by wide surgical resection and partial cystectomy.

Key Words: cystadenocarcinoma in situ, urachus, cystadenocarcinoma, cystadenoma

Introduction

The urachus is an embryological fibrotic remnant of the allantois, a canal that runs within the umbilical cord to drain the bladder of the fetus. The urachus remains patent in some adults throughout life. Adenocarcinoma of the urachus comprises less than 1% of bladder cancers, and it is mostly cystic and mucinous.[1] Adenomas are much rarer.[1] We describe a case of urachal mucinous cystadenocarcinoma in situ or cystadenoma of unknown malignant potential. We review the literature regarding the nomenclature, clinical behavior, management, and prognosis of this very rare condition.

Accepted for publication April 2012

Acknowledgement
The authors would like to thank Dr. Federica Balzarini for her help in translating the article by Palmieri et al.

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Case report

Patient and diagnosis

A 66-year-old man presented with long-standing history of lower abdominal pain and vague groin pain. He had no irritative lower urinary tract symptoms. He also had no hematuria or mucusuria. He denied any weight loss. His past medical history was unremarkable except for hypercholesterolemia and a benign cyst removal from his leg at the age of 16. Biochemistry and hematology investigations were within normal limits. Magnetic resonance imaging

(MRI) of his abdomen and pelvis revealed a large cystic mass compressing the dome of the bladder, Figure 1. The mass contained tiny calcifications as well as some loculations within the cyst. It was well defined but could not be separated from the anterior wall of the bladder at the dome. A flexible cystoscopy was done in the clinic. It revealed a depression on the anterior bladder wall at the dome secondary to external compression. At that same area, the mucosa was attenuated with whitish material underlying it, which raised suspicion for possible infiltration of the mass into the bladder itself. The presence of metastatic disease was excluded clinically and radiographically.

Surgery and follow up

The patient underwent surgical en-bloc resection of the mass with partial cystectomy and bilateral pelvic lymph node dissection. Intraoperatively, the mass was found to be arising from the urachus remnant. It was freely mobile with no peritoneal involvement. It was not involving the umbilical region. The urachus was identified and resected with the mass, but the umbilicus was spared. The mass, however, was inseparable from the dome of the bladder and a 2 cm bladder margin was removed with the specimen. Intraoperative frozen sections demonstrated that the surgical margins were negative for tumor. One year after the surgery, the patient was doing well with no signs and symptoms of recurrence.

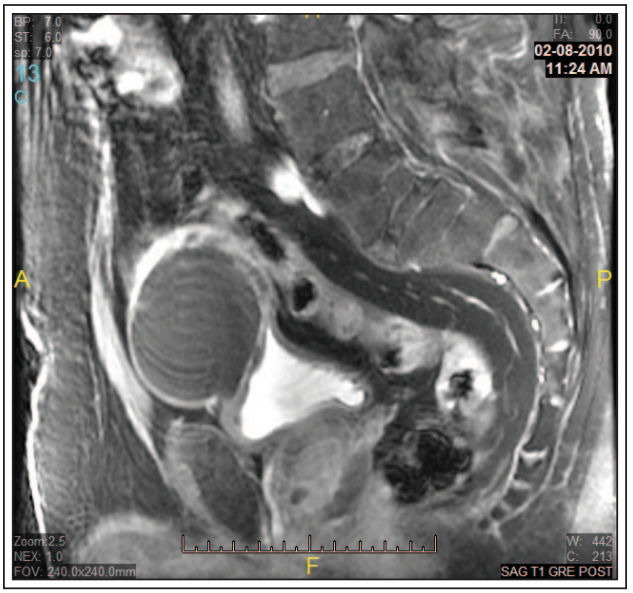

Figure 1. Contrast enhanced magnetic resonance imaging of the abdomen and pelvis shows a large cystic mass in the region of the urachus, well circumscribed compressing and inseparable from the bladder.

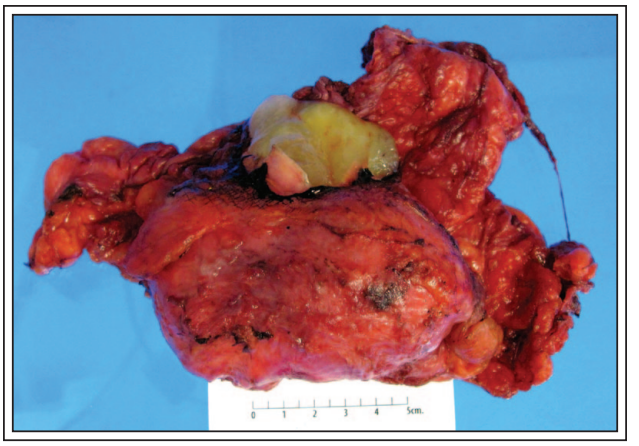

Figure 2. A large glistening mass measuring 9.0 cm x 8.0 cm x 6.0 cm. Incision of the tip of the cyst releases thick, yellow gelatinous material.

Pathology

The mass represented a unilocular cyst filled with thick, mucinous material and confined to the region of the urachal remnant, Figure 2. It measured 9.0 cm x 8.0 cm x 6.0 cm. The wall of the cyst was tan-yellow, with a thickness ranging from 0.1 cm to 0.9 cm, and contained multiple areas of calcification. On light microscopy, the cyst wall was predominantly lined by a single layer of cuboidal to columnar mucous cells characterized by pseudostratification, occasional papillary formation, and low grade cytologic atypia, Figure 3. No necrosis was seen. The cyst wall was fibrotic with dystrophic calcification. Occasional acellular mucus extravasation was seen within the fibrotic wall without epithelial cell component, but no peritoneal spread (pseudomyxoma peritonei) was seen.

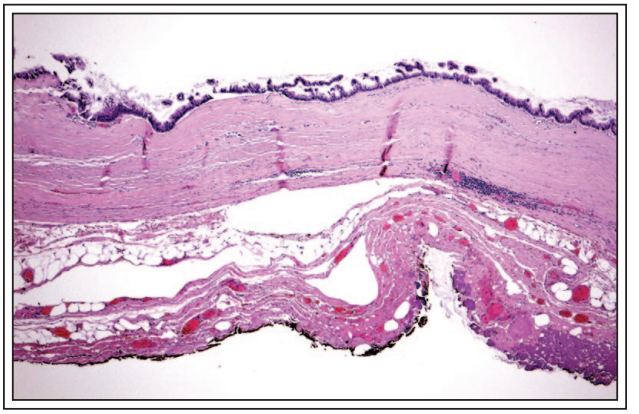

Figure 3. The cyst lumen is lined by a single layer of mucinous epithelium. No stromal invasion is present (Hematoxylin & eosin stain, x40)

TABLE 1. **Characteristics of reported cases of mucinous cystadenocarcinoma in situ or cystadenoma of unkown malignant potential**

Author/ year	Name	Size (cm)	Presentation	Gross			Pseudo-myxoma peritonei	Histology		Partial cystectomy
				Mucus filled	Unilocular	Communicating with bladder		Stromal invasion	Pleo-morphism/ atypia	
Hull and Warfe 1994	Adenoma §	14 x 8 x 5	Incidental finding	Yes	No	No	No	No	Yes	No
Paul et al. 1998	Adenocarcinoma in situ	3 cm*	Hematuria and mucusuria	Yes	Yes	Yes	No	No	Yes	Yes
Carr and McLean 2001	Adenoma of unknown malignant potential	4 x 3 x 3	Hematuria	Yes	No	No	No	No	Yes	Yes
Palmieri et al. 2002	Adenocarcinoma in situ	6.5 x 4.5	Heaviness sensation in hypogastric area and polyuria	Yes	No	No	No	No	Yes	No
Stenhouse et al. 2003	Adenocarcinoma in situ	14 x 9.5 x 7	Abdominal pain	Yes	Yes	No	Yes	No	Yes	—
Shinohara et al. 2006	Adenocarcinoma in situ	9 x 7 x 5.5	Inguinal hernia	Yes	Yes	No	Yes		Yes	Yes
Schell et al. 2009	Adenoma of unkown malignant potential	15.5x 8 x 8	Lower abdominal mass	Yes	No	No	No†	No	No	Yes
This case 2011	Adenocarcinoma in situ	9 x 8 x 6	Abdominal pain and groin pain	Yes	Yes	No	No†	No	Yes	Yes

§The tumor was only named as adenoma. However, pathology did show moderate pleomorphism.
†The patient did not have gross findings of pseudomyxoma peritonei. However, microscopic examination revealed mucous extravasation into perivesical soft tissue.
*Only a uni-dimensional measurement was provided by the authors.

Discussion

Mucinous epithelial tumors of the urachus that show cytologic atypia but lack the stromal invasion of an adenocarcinoma are very rare but can cause significant morbidity and mortality. Adverse clinical behavior can be either due to invasive growth and subsequent metastasis,[1] or due to rupture of the cyst with resulting pseudomyxoma peritonei, which can occur even in tumors without frank malignant cytology as pseudomyxoma peritonei is often related to ruptured low grade appendiceal mucinous neoplasm (mucinous cystadenoma).[2] A Medline literature review demonstrates that only seven similar cases have been reported to date. We summarize in Table 1, the clinical characteristics of all reported cases. Despite the two unique characteristics for all such tumors,

namely the presence of variable degrees of atypia and the absence of stromal invasion, there are two synonymous nomenclatures being used interchangeably. For tumors with no evidence of significant cytologic atypia such as nuclear pleomorphism, the term " low grade mucinous neoplasm of unknown malignant potential" is justified.[3] However, for all the others, we believe that the term "adenocarcinoma in situ" is a better description, as it reflects their potential to invade locally and metastasize. Accordingly, they should be treated similar to mucinous cystadenocarcinoma, which is a more studied entity with around few hundred cases reported in the English literature.[4] Several case series have studied urachal mucinous cystadenocarcinoma.[4-7] Such tumors have historically been associated with a poor prognosis, however, more contemporary series have demonstrated 5 year survival up to 40%.[4,6] These tumors are generally chemo-resistant, therefore the treatment is surgical with en-bloc resection along with partial cystectomy incorporating a wide excision margin.[8]

Ultrasound and computed tomography are the two imaging modalities generally used to assess urachal pathology.[9] MRI might be beneficial in identifying tissue invasion into the bladder[10] as well as patency of the urachus. Mucusuria and hematuria are signs that indicate invasion. However even in patients with cystadenocarcinoma, these are only present in 25% of cases.[9] Cystoscopy can be used to complement the assessment of bladder invasion, as in our case. Metastatic malignancy elsewhere should be ruled out clinically since the histologic diagnosis of in situ disease is frequently unknown at the time of clinical diagnosis. Most of the patients with the findings of cystadenocarcinoma in situ present initially with vague symptoms such as abdominal pain due to mass effect or with pseudomyxoma peritonei, Table 1. Although there is little supporting data, it may be serendipitous for such patients to have symptoms secondary to mass effect prior to the development of metastatic disease. After a complete diagnostic work up, we recommend treating these tumors as mucinous cystadenocarcinoma with wide surgical excision and close follow up of the patient. □

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