Mucinous cystic tumour of low malignant potential presenting in a patient with prior non-seminatous germ cell tumour

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Cite as: *Can Urol Assoc J* 2015;9(9-10):E750-3. http://dx.doi.org/10.5489/cuaj.2946
Published online October 13, 2015.

Abstract

Urachal neoplasms are rare entities which may be classified as cystic or non-cystic. Literature surrounding patient outcomes remains limited to non-cystic, urachal adenocarcinomas. Literature focusing on mucinous cystic neoplasms of the urachus is sparse. These mucinous cystic lesions may be subclassified as benign mucinous cystadenomas, mucinous cystic tumours of low malignant potential, and mucinous cystadenocarcinomas. Mucinous subtypes have the potential to behave aggressively and may result in pseudomyxoma peritonei. We describe here the case of a 37-year-old male with a mucinous cystic tumour of low malignant potential after prior right orchiectomy and left hydrocelectomy. This case raises the interesting possibility of multiple genitourinary neoplasms arising in a similar time frame.

Introduction

The urachus is an embryological structure that rarely persists in adults, but may be a site of malignancy. Any primary neoplasm of the urachus may be grouped as mucinous or non-mucinous, and as cystic or non-cystic.[1]

Mucinous cystic lesions may be subclassified as benign mucinous cystadenomas, mucinous cystic tumours of low malignant potential, and mucinous cystadenocarcinomas. Mucinous subtypes have the potential to behave aggressively and may result in pseudomyxoma peritonei (PMP).[2] However, natural progression of these lesions is difficult to interpret given the lack of reports. We attempt to further this understanding by describing the case of a 37-year-old male who presented with a mucinous cystic tumour of low malignant potential after prior right orchiectomy.

Case report

This 37-year-old male patient originally underwent right radical orchiectomy for a stage pT2, non-seminomatous germ cell tumour (NSGCT) (95% embryonal, 5% seminomatous) with lymphovascular invasion and elevated lactic acid dehydrogenase in early 2013. During workup, computed tomography (CT) demonstrated a 3.5-cm cystic, non-enhancing lesion with calcification contiguous with the right upper aspect of the bladder (Fig. 1). Cystoscopy demonstrated no bladder connection. This mass was determined to be unrelated to the testicular tumor given its appearance and lack of retroperitoneal lymphadenopathy. There were no pulmonary, hepatic, or skeletal metastases. Following orchiectomy, the patient underwent 2 cycles of adjuvant BEP (bleomycin, etoposide, cisplatin) chemotherapy with no recurrence of testicular tumour.

Eighteen months later, the patient underwent left varicocelectomy for a varicocele, but developed a left hydrocele as sequelae. It was decided that the urachal mass be removed at the time the hydrocele was addressed. The patient then underwent concurrent left hydrocelectomy and partial cystectomy. At time of surgery, there was no evidence of PMP.

Pathology findings

Gross examination revealed a cystic structure containing cloudy, mucinous material that measured 4.0 cm in diameter with a wall thickness of 0.2 cm. Microscopic sections through the bladder resection revealed a thin-walled fibrous diverticulum (Fig. 2). The inner wall of the diverticulum contained granular calcified debris eliciting a multinucleated giant cell reaction. An area of intact mucosa was composed of columnar epithelium showing mucinous metaplasia with papillary architecture. Mucin was present in the lumen of the diverticula and had extravasated into the wall. The tumour was positive for cytokeratin 7 and cytoplasmic beta catenin. There was no invasive component. A nodule of smooth

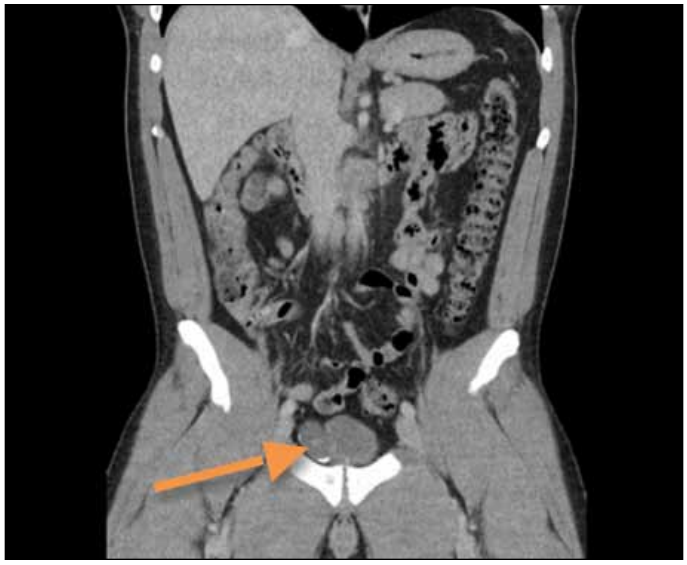

Fig. 1. Computed tomography scan demonstrating an irregular cystic lesion with calcification contiguous with the right anterior upper aspect of the bladder.

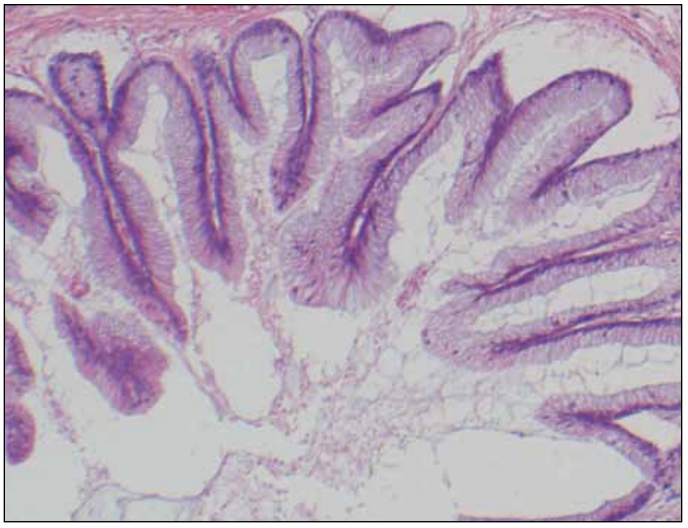

Fig. 2. Pathology specimen through the bladder resection demonstrating a thin walled fibrous diverticulum with abundant inner wall granular calcified debris, a small area of intact mucosa showing mucinous metaplasia, and luminal mucin extravasating into the wall. No invasive components were noted.

muscle contained cystic spaces lined by urothelium, a feature diagnostic of urachal remnant (Fig. 3). It was determined that the lesion was consistent with a diagnosis of mucinous cystic tumour of low malignant potential (MCTLMP).

Discussion

Glandular tumours of the urachus remain sparsely described due to rarity, degree of clinical correlation required to establish urachal origin, and lack of standardized nomenclature. A recent classification scheme was proposed by Amin and colleagues,[1] assessing 55 cases at their institution. Of these, 22 were classified as MCTLMP. There are 5 other reports of lesions consistent with MCTLMPs.[3-7] (Table 1).

Other mucinous cystic lesions of the urachus include mucinous cystadenoma and cystadenocarcinoma.[8-10] Mucinous cystadenomas display no dysplastic change, MCTLMPs demonstrate dysplastic change without invasion, and mucinous cystadenocarcinomas demonstrate invasion. It is hypothesized that mucinous cystadenomas and MCTLMPs represent a continuum of lesion which may eventually degenerate into mucinous cystadenocarcinoma. MCTLMPs are managed surgically, although there is no evidence that definitively demonstrates the degeneration of MCTLMPs into mucinous cystadenocarcinoma. Adults suspected of having a urachal remnant of any sort may be at high risk of malignancy.[11] Predictors of urachal malignancy include patients over 55 and patients with hematuria. Prophylactic excision of urachal anamolies in children is not recommended. Many patients likely undergo treatment to prevent a single case of urachal adenocarcinoma.[12]

MCTLMPs are most often incidental findings, although patients may present with an abdominal mass, pain, mucusuria, hematuria, urgency, or umbilical discharge. Patients may also demonstrate PMP originating from the urachal mass. Two prior cases of MCTLMP have demonstrated PMP,[5,6] which is associated with complications such as cachexia and bowel obstruction if left untreated. Current treatment of PMP includes total urethrectomy, partial cystectomy, and peritonectomy followed by adjuvant chemotherapy.[13]

Our patient had a significant history of urologic issues, namely a right-sided, high-risk NSGCT and left varicocele. No other cases of MCTLMPs with prior urologic history have been documented. This patient's NSGCT was primarily embryonal with positive lymphovascular invasion, stratifying it as high risk with up to a 50% chance of recurrence.[14]

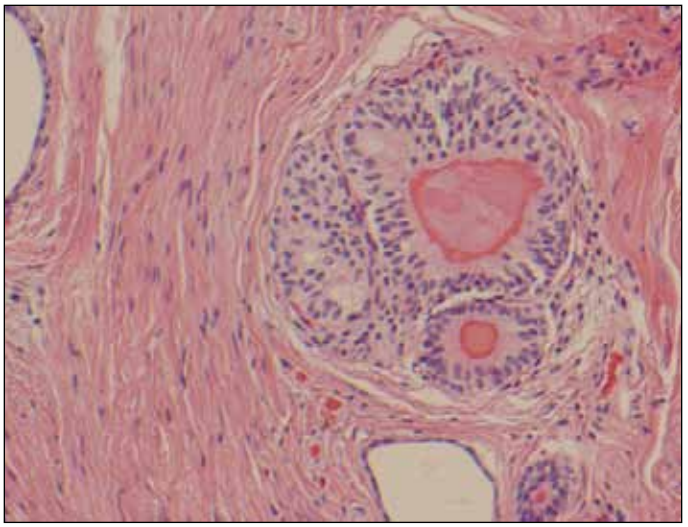

Fig. 3. Pathology specimen containing a nodule of smooth muscle containing cystic spaces lined by urothelium; features diagnostic of urachal remnant.

Table 1. Cases of MCTLMP or similar lesions thus far reported in case reports

Reference	Age	Sex	Size (cm)	PMP	Diagnosis	Treatment	Symptoms	Patient history
Present case	37	M	4	Absent	MCTLMP	Partial cystectomy concurrently with left hydrocelectomy	Incidental finding	Prior right radical orchiectomy, prior left varicocelectomy
Carr and McLean[3]	72	M	4	Absent	Mucinous tumour of uncertain malignant potential	Partial cystectomy	Hematuria (microscopic), nocturia	No other medical issues
Paul et al[4]	68	M	3	Absent	Stage 0 mucinous adenocarcinoma in situ	Partial cystectomy	Hematuria, mucusuria	No other medical issues
Shinohara et al[5]	54	M	6	Present	Mucinous cystic tumour with low malignant potential	Partial cystectomy	None	Mucus from ruptured urachal cyst found during left inguinal hernia repair, cyst excised at re-operation
Stenhouse et al[6]	54	M	11	Present	Mucinous neoplasm of uncertain malignant potential	N/A	Abdominal pain, rectal bleeding	No other medical issues
Choi et al[7]	29	F	5.5	Absent	Mucinous tumour of uncertain malignant potential	Partial cystectomy	Right flank pain	No other medical issues

MCTLMP: mucinous cystic tumour of low malignant potential; PMP: pseudomyxoma peritonei; N/A: not available; M: male; F: female.

Table 2. A case series of MCTLMP or similar lesions

Reference	Age	Sex	Size (cm)	Diagnosis	Treatment	Symptoms
Amin et al[1]	48	F	8	MCTLMP with intraepithelial carcinoma	Excision of urinary bladder mass & umbilectomy	Suprapubic and umbilical mass
	26	F	2	MCTLMP	Partial cystectomy	Suprapubic mass
	74	M	6.5	MCTLMP	Excision of tumour & sigmoid colectomy	Incidental finding
	72	M	0.8	MCTLMP	Partial cystectomy	Mucusuria
	74	M	3	MCTLMP	Partial cystectomy	Hematuria
	50	F	2.1	MCTLMP	Resection of urachus	Mass
	45	M	3.5	MCTLMP	Partial cystectomy	RLQ pain, Hematuria
	58	F	1	MCTLMP	Excision of lesion	Incidental finding
	43	F	2.5	MCTLMP	Partial cystectomy	Incidental finding
	40	F	6	MCTLMP	Partial cystectomy, urachectomy, and umbilectomy	Incidental finding
	80	F	2.5	MCTLMP	Partial cystectomy and urachectomy	Mucusuria
	37	F	N/A	MCTLMP	N/A	Incidental finding
	29	F	N/A	MCTLMP	N/A	Bladder dome nodule
	N/A	N/A	N/A	MCTLMP	N/A	N/A
	42	F	8	MCTLMP	Excision of pelvic mass	Pelvic mass
	42	F	6	MCTLMP	N/A	Midline cystic mass
	36	F	N/A	MCTLMP	N/A	Incidental finding
	39	M	6.5	MCTLMP	Umbilectomy and urachectomy	Obstruction and umbilical discharge
	57	M	2.8	MCTLMP with intraepithelial carcinoma	Partial cystectomy	N/A
	77	F	5.5	MCTLMP	Partial cystectomy	N/A
	43	M	7	MCTLMP	Partial cystectomy, umbilectomy, and urachectomy	Incidental finding
	26	M	8	MCTLMP	Partial cystectomy	Urgency, abdominal pain

MCTLMP: mucinous cystic tumour of low malignant potential; RLQ: right lower quadrant; N/A: not available; M: male; F: female.

Although there is debate regarding post-surgical management of high risk NSGCT, options include 1 to 2 cycles of adjuvant BEP or surveillance.[15] If a tumour recurs during surveillance, 3 to 4 cycles of BEP may be required. Our patient was well-advised about these options and opted for adjuvant BEP.

Additionally, one-fifth to two-thirds of patients with tumours composed primarily of embryonal carcinoma have metastases at presentation, highlighting importance of ruling out this possibility.[16] Lack of connection to the bladder, non-enhancement during imaging, and no evidence of retroperitoneal lymphadenopathy or other metastases suggested the lesion was unrelated to the testicular tumour. It was opted that surgical excision of this lesion wait until the patient had undergone complete treatment of his testicular cancer. This is a challenging clinical decision given the lack of data on possible malignant degeneration of urachal neoplasms.

We suspect our patient had underlying genetic predispositions due to his testicular and urachal neoplasms, and his young age at MCTLMP presentation. Monitoring this patient for recurrence or new lesions is warranted.

Conclusion

We have presented the case of a patient presenting with a urachal MCTLMP in the context of treatment for a right-sided testicular NSGCT. Clinical assessment of MCTLMP remains difficult and may present challenge in terms of timing of surgery. Lack of data on the natural history of these lesions and their possible malignant degeneration warrant that MCTMKP be treated with partial cystectomy.

Competing interests: The authors declare no competing financial or personal interests.

This paper has been peer-reviewed.

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