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Case Report
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Endoscopic Management of Urachal Mucinous Cystic Tumor of Low Malignant Potential (MCT-LMP)

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Abstract

Urachal mucinous tumors are rare neoplasms with behavior that can range from relatively benign to malignancy that can spread distantly or throughout the peritoneum (pseudomyxoma peritonei or peritoneal carcinomatosis). Here we describe a case of a small (2 cm) urachal Mucinous Cystic Tumor of Low Malignant Potential (MCT-LMP), confined to an intact cyst at the dome of the urinary bladder, without rupture or peritoneal spread. The urachal mucinous tumor was an incidental finding on a staging CT-scan performed for nonspecific gastrointestinal disorder. The cystic lesion at the dome of the bladder was totally resected by Transurethral Resection of the Bladder (TURB). No additional surgery, systemic chemotherapy or radiation therapy following TURB was done. Thirty months after surgery, the patient had no evidence of metastasis or local recurrence. This case raises the interesting possibility of endourology conservative management for small urachal MCT-LMP, with no recurrence and metastases after intermediate-range follow-up.

Keywords: Urachus; Urachal adenocarcinoma; Mucinous cystic neoplasm

Introduction

Urachal neoplasms are a rare and well-known subset of bladder tumors arising from neoplastic transformation of remnant urachal tissue [1-5]. With diverse clinical presentations and varied gross and microscopic features, most urachal neoplasms are glandular neoplasms, typically with an intestinal phenotype [6-8]. A recent classification terminology was proposed using two broad categories: (1) tumors that were non-cystic and frankly invasive; (2) tumors having a prominent cystic component, appearing overall low grade and showing a remarkable homology to mucinous cystic tumors of the ovary [6]. In contrast to the non-cystic adenocarcinoma which mostly presents with hematuria, the most frequent cystic urachal lesions diagnostic was incidental or related to a mass lesion (each 32%), followed by hematuria (24%), mucusuria (12%), or pain (12%) [6]. Among reported urachal glandular neoplasm, primary urachal cystic tumors are exceedingly rare, and in this spectrum, 65% have been classified as urachal Mucinous Cystic Tumor of Low Malignant Potential (MCT-LMP) [6,9]. MCT-LMP have only been previously studied in small series and individual case reports, and their prognosis is not clearly established. However, it has been shown that progression free survival of non-invasive mucinous cystic tumors is significantly better than non-cystic invasive adenocarcinoma [6,8-10]. Cross-sectional techniques (multiplanar CT-Scan or MRI imaging) are the best way for diagnosis, midline supravesical location is a key feature [11], and calcification occurs in 50% to 70% [12].

Histology of urachal MCT-LMP may show areas resembling mucinous cystadenoma but also has stratified cyst lining of usually no more than 3 cell layers. Mucin extravasation into the cyst wall has been described, often with calcification or rarely ossification. Immunohistochemical characterization that did not differ significantly between cystic and non-cystic tumors (CK7+, CK20+, CDX2+, b-catenin-, ER-, and PR-) [13], was reported distinct from that of colonic adenocarcinomas (CK7- and b-catenin+ (nuclear)) [6,13].

Most MCT-LMP have been treated by aggressive surgery (*i.e.* partial or radical cystectomy), due to their size at diagnosis, and the potential to behave aggressively (resulting in recurrence and pseudomyxoma peritonei) [6,14,15]. Rare cases have been managed by conservative treatment (5%) (i.e. Transurethral Resection of Bladder tumor (TURB)) [6,15]. Primary treatment strategy for cystic

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tumors deemed to be urachal is partial cystectomy, urethrectomy, and umbilectomy (with good clinical behavior), whereas the treatment for primary adenocarcinoma of the bladder mucosa is radical cystectomy [2,4,6,9,15,16]. Indeed, suboptimal management can lead to peritoneal spread and intraabdominal tumor implants. Therefore, these neoplasms are often managed with complete surgical excision as proper surgical management [6,17], even if some emerging controversy exist on partial *vs.* complete cystectomy [1,9,18]. Little consensus for the role of adjuvant or neoadjuvant chemotherapy or radiation therapy in urachal adenocarcinoma have been done. Some evidence came out with cisplatin and 5-fluorouracil based regimens recently [6,19]. We attempt to further this understanding by describing this new case of a 70-year-old male who presented with a small urachal mucinous cystic tumor of low malignant potential lucky find. This case raises the interesting possibility of conservative management (TURB) for small urachal MCT-LMP with no recurrence and metastases after 30 months intermediate-range follow-up, confirming good outcomes [6,9].

Case Presentation

We describe the case of a 70-year-old male retired firefighter referred for investigation of an incidental abdominal mass abutting the dome of the bladder. It was identified initially on abdominal CT scan as part of an investigation for nonspecific gastrointestinal disorder. The patient reported lower urinary tract symptoms as nocturia and pollakiuria without hematuria. Data regarding clinical history, diagnostic imaging, and pathology were collected retrospectively. His past medical history was unremarkable except an operated inguinal hernia. Care was provided at a tertiary care teaching hospital and the patient provided written consent for a review of medical records and for publication of a case report, in accordance with institutional policy. During workup, multiphasic contrast-enhanced abdominopelvic Computed Tomography (CT) demonstrated 2 cm low-attenuation nodular thickening of the anterior bladder wall with peripheral calcifications without enhancement in either nephrographic or excretory phase, suspected to represent a bladder diverticulum or a urachal cyst (Figure 1). Cystoscopy demonstrated submucosal deformation (2 cm long axis) with a tumor sluice and per endoscopic mucus expelling (Figure 2). The cystic lesion at the dome of the bladder was totally resected (TURB) and sent as two specimens (superficial and deep) to pathology. At the time of surgery, there was no evidence of pulmonary, hepatic, or skeletal metastases, and pseudomyxoma peritonei or peritoneal carcinomatosis. There were no postoperative complications. Microscopic sections through the bladder resection revealed the epithelial lining consisted of a single layer of cuboidal to columnar epithelial cells with an intestinal phenotype, including scattered goblet cells and focal low-grade dysplasia in urachal remnants. The lumen of the cyst contained acellular mucin, which dissected in some areas into the partially calcified cyst wall, but did not reach the serosal surface (Figure 3). In our case report, there was no invasive component and immunohistological characterization was not done.

Then the cyst was felt to be best classified as a urachal mucinous cystic tumor of low malignant potential, based on the classification system described by Paner et al. [8]. The patient did not receive additional surgery, systemic chemotherapy or radiation therapy following TURB. He was followed each 6 months by cystoscopy and additional MRI. Thirty months after surgery, the patient had no evidence of metastasis or local recurrence.

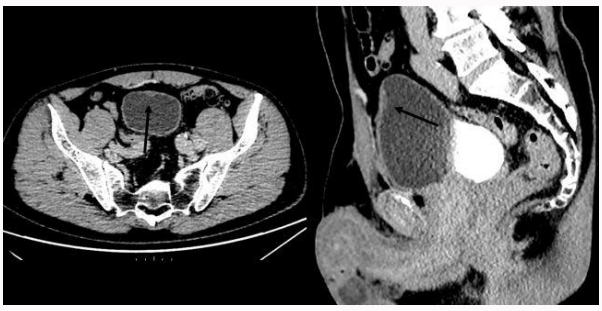

Figure 1: Multiphasic contrast-enhanced abdominopelvic Computed Tomography (CT) demonstrated 1.5 cm low-attenuation nodular thickening of the anterior bladder wall with peripheral calcifications without enhancement in either nephrographic or excretory phase. (black arrow; axial and sagittal views).

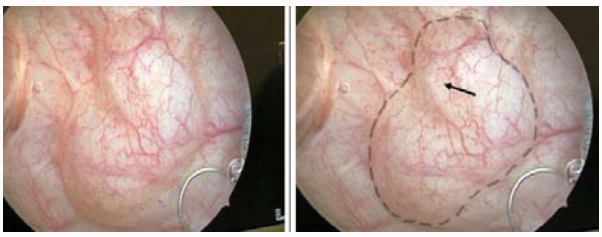

Figure 2: Endoscopic intraoperative image. Submucosal deformation (2 cm long axis; black dot marking). Tumor sluice with per endoscopic mucus expelling (black arrow).

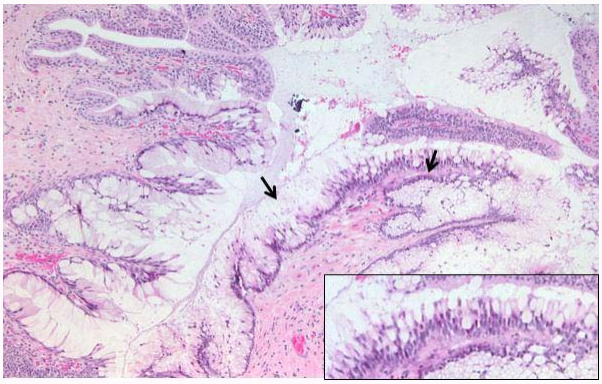

Figure 3: Histology of urachal MCT-LMP; cyst epithelial lining with goblet-cell (black arrow) and focal low-grade dysplasia (high magnification) in urachal remnants (top).

Conclusion

The urachus may be the site of glandular tumors of varying morphologic appearances and behavior. These tumors may be cystic mucinous neoplasms, which resemble ovarian mucinous cystic tumors and may be benign, of low malignant potential, or frankly malignant. Our case report reveals the feasibility of a conservative management (TURB) without invasive abdominal wall mass resection for small sized urachal Mucinous Cystic Tumor of Low Malignant Potential (MCT-LMP).

Classification between cystic and non-cystic and tumor size of urachal tumors could be an informative part for prognostic and therapeutic significance supporting decisions for conservative management. However, caution is especially advised when dealing with low grade mucinous tumors, particularly in limited material.

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