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Oncology

Myoepithelial carcinoma of the bladder: Case report and review of the literature

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ABSTRACT

Myoepithelial carcinoma is a neoplasm that classically arises in the parotid glands, nasopharynx, paranasal sinus, and nasal cavity of the head and neck. It rarely arises in other organs or soft tissues and involvement of genitourinary organs is distinctly rare. We describe a case of a 21-year-old male, presenting with nausea, weight loss, and worsening suprapubic pain over 3 months, found to have a large mass at the dome of the bladder. Partial cystectomy was ultimately performed revealing myoepithelial carcinoma of the bladder. The patient is free of disease at four years without the need for systemic therapy.

1. Introduction

Bladder cancer commonly arises from urothelial cells and mesenchymal neoplasms arising from the connective tissue of the bladder are unusual, typically originating from smooth muscle or rarely other cell lineages. Myoepithelial cells are usually found in glandular epithelium. Neoplasms arise from transformation of these cells in organs where they normally reside; however, rare *de novo* tumors have been described arising in other organs.[1–3] Histopathologic features can be variable due to morphologic diversity, involving spindle, plasmacytoid, epithelioid, or clear cell types with one cell type usually predominating. To confirm these diverse lesions are true myoepithelial neoplasms, immunohistochemistry is performed to show expression of cytokeratins and one or more markers for S-100 protein, calponin, p63, GFAP, maspin, and actin. Myoepithelial carcinoma (MC) of the bladder is exceptionally rare. Here we describe our experience with this disease in a single patient and review the existing literature on genitourinary MC.

2. Case presentation

A 21-year-old man with diabetes presented to our emergency department with 3 months of worsening suprapubic pain and weight loss. Outside imaging showed a 5cm mass at the bladder dome, biopsy of which was inconclusive. Repeat imaging at our facility redemonstrated the heterogeneous mass (Fig. 1A), and cystoscopy confirmed a large, ulcerated lesion without other sites of disease (Fig. 1B). As the biopsy was negative for adenocarcinoma and imaging showed no extravesical disease or urachal involvement, decision was made to proceed with robotic partial cystectomy with pelvic lymph node dissection.

Pathology revealed a 4.5cm tumor based in the muscularis propria composed of epithelioid and spindled cells with prominent nucleoli invading between bundles of muscularis propria (Fig. 2). Vascular invasion and focal necrosis were present. No urothelial or other neoplasm was present. Margins and nodes were free of disease. Given the unusual cytology, a broad panel of immunohistochemical stains were performed to classify the tumor lineage. Markers associated with Ewing sarcoma, rhabdomyosarcoma, melanoma, and additional epithelial markers were negative. MC was deemed the most consistent interpretation; the case was reviewed by an extramural expert consultant with the same interpretations rendered.

Postoperative bone scan and chest x-ray were negative for metastasis. Incidentally, the patient developed episodic urinary retention requiring intermittent catheterization. Urodynamic testing indicated

Abbreviations: GFAP, Glial fibriraly acidic protein; MC, Myoepithelial Carcinoma; SMA, Smooth muscle actin; UCC, Urothelial Cell Carcinoma; TREP, Tumori Rari in Etá Pediatrica.

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Abbreviations

GFAP	Glial fibrillary acidic protein
MC	Myoepithelial Carcinoma
SMA	Smooth Muscle Actin
UCC	Urothelial Cell Carcinoma
TREP	Tumori Rari in Etá Pediatrica

include papillomas and adenomas, while neurofibromas and hemangiomas are benign nonepithelial tumors. In children under 10, the most common cause of a bladder cancer is rhabdomyosarcoma, and less commonly neuroblastoma, undifferentiated sarcoma, and primitive neuroectodermal tumors.

MC is rare and characterized by variable cellular morphology. The cytologic appearance is also variable in cellularity and background stromal quality. Due to this heterogeneity, demonstration of co-expression of epithelial, neural, and smooth muscle markers is often necessary for diagnosis.[4] Cases of MC are most commonly associated

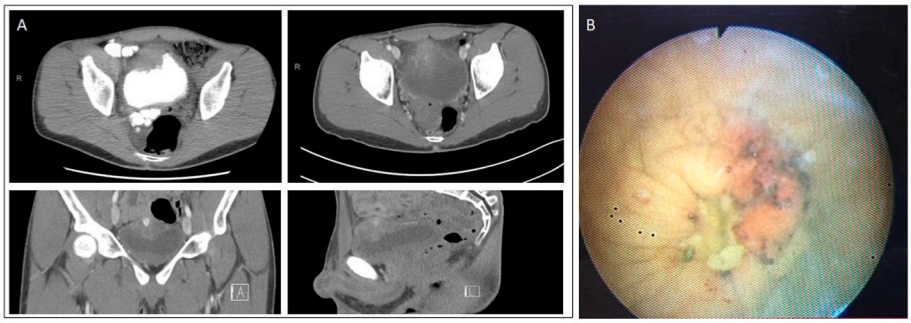

Fig. 1. CT and cystoscopic imaging of bladder mass. A) CT cystogram with transverse image, and CT with IV contrast with transverse, coronal, and sagittal images. B) Cystoscopy showing large ulcerating mass at the dome of the patient's bladder.

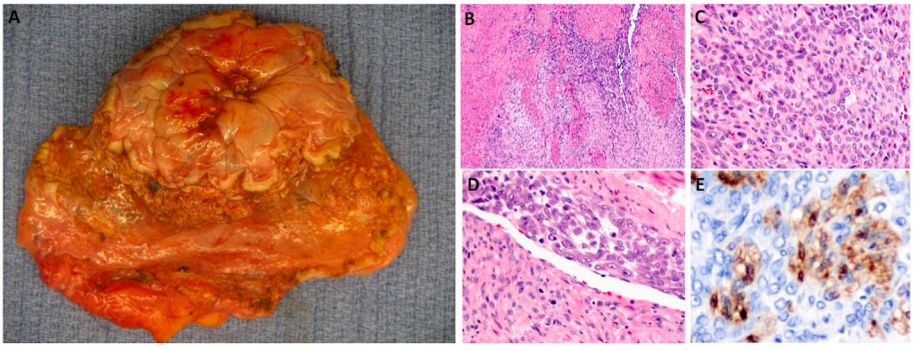

Fig. 2. Pathological specimen following partial cystectomy. Gross pathology of the 4.5cm partial cystectomy specimen, here photographed from the luminal aspect (A). Histopathology of bladder myoepithelial carcinoma at low power revealing an infiltrative cellular neoplasm invading between bundles of muscularis propria smooth muscle (B). At high power, variable epithelioid to spindle cytomorphology is evident, as are prominent nucleoli (C). Among worrisome features seen, foci of vascular invasion were identified, with a plug of carcinoma filling an endothelial-lined vascular space (D). The unusual immunophenotype seen was the pattern characteristic of myoepithelial neoplasms, including coexpression of pancytokeratin AE1/AE3, cytokeratin CAM5.2, S100, and Calponin (E).

decreased sensation with poor compliance and bladder outlet obstruction, thought to be multifactorial related to his poorly controlled diabetes, surgically reduced bladder capacity, and dysfunctional voiding. The case was reviewed at our institution's Interdisciplinary Genitourinary Tumor Board conference, and no adjuvant therapy was recommended. He was followed without evidence of disease recurrence for the first three years. In year four, he presented to the ER with acute abdomen requiring exploratory laparotomy, with a finding of a small perforation at the dome of the bladder. Repeat partial cystectomy was performed at this location, with pathology notable for nephrogenic adenoma but no disease recurrence. We plan to continue annual surveillance until he is five years free from recurrence, with closer follow up for his neurogenic bladder.

3. Discussion

The differential diagnosis for bladder tumors is broad and includes both benign and malignant etiologies. Most primary bladder tumors are epithelial and include malignant carcinoma in situ, urothelial cell carcinoma (UCC), squamous cell carcinoma, and adenocarcinoma. Malignant tumors of nonepithelial origin include leiomyosarcoma, lymphosarcoma, and chondrocarcinoma. Benign epithelial tumors

with the head and neck but have also been described as arising from the breast, lung, and liver. Urological sites of origin are extremely rare, but MC has been reported in the scrotum and bladder.[1–3] In our case, MC was diagnosed by appropriate morphology in context of expression of epithelial, neural, and smooth muscle markers, consistent with established diagnostic criteria. In this context, focal necrosis, vascular invasion, and prominent nucleoli additionally indicated malignancy.

Management is characterized by complete excision of the mass ±perioperative chemoradiation. MC is generally a low-grade malignant tumor with 23–50% local recurrence, so routine follow-up is required.[1] Rarely, this neoplasm may spread via lymph nodes or hematogenous metastasis. After initial excision of the tumor, adjuvant chemotherapy or radiotherapy has been reported with mixed results.

The first report of primary bladder MC was described in 2016 in a 61-year-old woman presenting with hematuria. The patient was treated with primary cystectomy. Metastases were identified within 2 years.[3] The second report of bladder MC is in a pediatric patient who also presented with hematuria. This patient was treated with neoadjuvant ifosfamide, cisplatin, and etoposide chemotherapy, surgical resection with partial cystectomy, and adjuvant radiation as suggested in the Tumori Rari in Etá Pediatrica (TREP) project aimed at developing management strategies for rare childhood solid malignancies.[2,5] Our

approach to this tumor—partial cystectomy with nodal dissection—is analogous to treatments conducted in these reports and elsewhere in the body. Pediatric MC may be more aggressive than similar malignancies presenting in adulthood.[2] Our patient presented at age 21 and may have had tumor progression since his late teens making this a potential extension of a pediatric malignancy into early adulthood. As such, we continue to monitor our patient for disease recurrence and management of his neurogenic bladder.

4. Conclusion

UCC is the most common form of bladder cancer. MC typically arises from sites in the head and neck and primary GU MC is rare. To our knowledge, we present the first case of adult bladder MC treated by partial cystectomy and followed by surveillance without recurrence to date. There is currently no standard treatment for genitourinary MC. The recommendation for treatment in other regions is complete resection ±chemoradiation. Future research is needed to identify the optimal treatment modality for these malignancies.

Consent

The patient provided written informed consent to publish the details of their medical case, including publication of images. This study was reviewed by the Virginia Commonwealth University Office of Research and Innovation Institutional Review Board and was found to be not classified as human subjects research.

Author contributions

Dr. Bednarz, Student Doctor Lanyi, and Dr. Wilson contributed to background research, manuscript drafting and revision. Dr. Smith reviewed the pathology and contributed to the manuscript. Dr. Hampton contributed to the patient's medical management and manuscript revision. Dr. Krzastek contributed to the patient's medical management, manuscript design and revision.

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Data availability statement

All data generated or analyzed during this study are included in the article. Further enquiries can be directed to the corresponding author.

Declaration of competing interest

The authors have no disclosures.

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