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Human PATHOLOGY Case Reports

## Case Report

## Primary urinary bladder angiosarcoma with ascites

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#### ABSTRACT

Non-urothelial tumors of the urinary bladder are rare. Sarcomas constitute less than 1% of all malignant neoplasms and angiosarcomas comprise 2% of all sarcomas [1]. We report a case of a 69-year old male with history of a low-grade prostatic adenocarcinoma for which he underwent external beam radiation therapy in 1/2010. He presented to the ER in 9/2015 with an abdominal distension and urinary retention and was found to have malignant ascites, hydronephrosis and a urinary bladder mass. Pathologic evaluation of the ascites and urinary bladder tumor revealed metastatic epithelioid angiosarcoma. Since the patient presented with an advanced stage inoperable disease, chemotherapy was initiated shortly thereafter. Unfortunately, the patient became septic and died 6 weeks following the original diagnosis.

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#### 1. Introduction

Primary angiosarcoma arising in the urinary bladder is an exceedingly rare malignant neoplasm. To our knowledge, there are less than thirty reported cases in the published literature [1–15]. The first case of urinary bladder angiosarcoma reported in literature dates back to 1907 [6] and the largest series of epithelioid angiosarcomas (nine cases) was reported by Matoso and Epstein [5].

Overall, soft tissue sarcomas comprise less than 1% of all malignant neoplasms. Angiosarcomas comprise 2% of all soft tissue sarcomas. The majority of the angiosarcomas occur in the head and neck area [7]. Visceral organ involvement of angiosarcoma is less common. In this report, we describe the clinical presentation and pathologic features of a patient with an epithelioid angiosarcoma of the urinary bladder.

## 2. Case report

### 2.1. Clinical presentation

A 69-year old male presented to an Emergency Department with a chief complaint of abdominal distension for one week. Past medical history was significant for an adenocarcinoma of the prostate, Gleason score 3+3=6, involving two cores (left body, right body) in an extended sextant biopsy with a maximal linear extent of 1 mm diagnosed in January, 2010. He had a prior prostate biopsy seven weeks earlier which contained two cores with high-grade prostatic intraepithelial

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neoplasia (left base and left body). The patient underwent external beam radiation therapy between 04/12/2010 and 06/11/2010. During this time, he received 75.6 Gy in 42 fractions. Aside from radiotherapy, patients with low risk prostate disease can pursue active surveillance or radical prostatectomy as their treatment options. Our patient did well until September 2015, when he developed abdominal distension associated with acute renal failure and urinary retention. A CT scan of the abdomen and pelvis showed a large amount of ascites, right side hydronephrosis (Fig. 1A) and a large area of asymmetric posterior and right lateral urinary bladder wall thickening (up to 1.7 cm in thickness) (Fig. 1B). Cystoscopy revealed bladder neck narrowing with ulceration and whitish tissue in the posterior and right lateral walls. Transurethral resection of the abnormal bladder wall tissue was performed. The patient also underwent paracentesis of the ascites.

### 2.2. Pathology

#### *2.2.1.* Urinary bladder (transurethral resection)

Grossly, the bladder tissue consisted of numerous pink-tan soft tissue fragments measuring 2.9 cm  $\times$  2.5 cm  $\times$  1.0 cm in aggregate. Histologic examination revealed a poorly differentiated malignant neoplasm extensively infiltrating the bladder lamina propria and the detrusor muscle. No prostatic glands were identified. Two morphological architectural patterns were identified. The more prevalent pattern was comprised of tightly packed sheets of intermediate size round to oval neoplastic cells. The other pattern was characterized by luminal formation with abortive vascular structures lined by similar appearing neoplastic cells without intraluminal cells or other material.

The neoplastic cells had a predominantly "epithelioid" to focally slightly spindled appearance, characterized by cells with high nuclear

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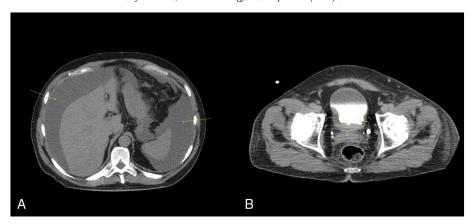


Fig. 1. CT scan images of angiosarcoma. (A) Noncontrast axial CT scan through the upper abdomen demonstrates low density ascites filling the peritoneal cavity. (B) CT scan of urinary bladder tumor. An axial image from an IV contrast enhanced CT urogram shows an irregular mass causing thickening of the right lateral, posterior, and left posterolateral bladder wall.

to cytoplasmic ratios, hyperchromatic nuclei, variably prominent nucleoli and focal nuclear molding (Fig. 2). A comprehensive immunohistochemical panel was performed. The neoplastic cells were diffusely immunoreactive for vimentin and endothelial cell markers (CD31, von

Willebrand factor and ERG) (Fig. 3) and were negative for epithelial markers (cytokeratin AE1/AE3, cytokeratin CAM 5.2, cytokeratin 903, and epithelial membrane antigen), prostatic markers (prostate specific antigen (PSA), androgen receptor prostein (P501S) and NKX3.1),

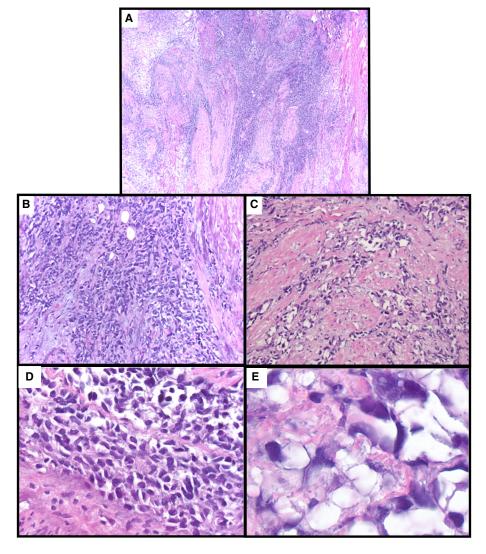


Fig. 2. Histopathologic features of the urinary bladder tumor: A) Low magnification image showing a cellular neoplasm infiltrating deeply into the detrusor muscle ( $H \& E, 40 \times$ ). B) Tumor cells exhibit sheet-like arrangement of epithelioid and spindle cells ( $200 \times$ ). C) Tumor cells show abortive vessels with lumen formation ( $200 \times$ ). D) Abortive vessels ( $400 \times$ ). E) The tumor cells delineate epithelioid features ( $400 \times$ ).

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