Malignant Hemangiopericytoma of the Bladder: Case Report and Literature Review

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Abstract

Hemangiopericytoma/Solitary Fibrous Tumor (HPC/SFT) of the bladder is a very uncommon neoplasm of uncertain malignant behavior. 10 previous cases of HPC/SFT in the bladder have been reported in the literature. Only two cases have been malignant. In this manuscript, we present the third malignant HPC/SFT of the urinary bladder and also review the literature for this very rare tumor.

Keywords: Bladder Cancer; Cystectomy; Literature review; Malignant Hemangiopericytoma; Solitary Fibrous Tumor

Introduction

Bladder cancer is the second most common malignancy of the urogenital system with over 74,000 new cases diagnosed each year [1]. Urothelial/transitional cell carcinoma is the predominant histologic subtype, accounting for 90% of all bladder cancers in the US and Europe [1]. Conversely, we identified only 10 previous cases of bladder HPC/SFTs. Malignant cases are even less common with only two documented cases. In 2002, the world's soft tissue tumor experts reclassified HPC under soft tissue tumors attributing to its fibroblastic rather than pericytic origin [2]. First described in 1942 by Stout and Murray, HPC has been found throughout the body and makes up of 2.5% of all soft tissue tumors [3,4]. In the genitourinary system, the kidney has the highest prevalence of HPC/SFTs (approximately 50 reports) with fewer cases noted in the prostate and ureter [5,6,7]. Although HPC has been associated with a history of trauma, prolonged steroid use, and hypertension, a definitive correlation does not exist [8]. The rarity of this bladder mass makes its behavior unpredictable and treatment and clinical follow up remain uncertain. This report presents a case of bladder HPC/SFT and provides a literature review.

Case Report

A 56 year-old African American male with a history of hypertension, chronic back pain, and alcoholism presented with 2 episodes of painless gross hematuria followed by urinary

retention. A urine culture showed no growth. No cytology specimen was obtained. A contrasted Computed Tomography (CT) scan was obtained and demonstrated an enlarged prostate with a prominent median lobe and evidence of an intravesical bladder neck mass without lymphadenopathy (Figure 1).

Follow up cystoscopy clearly showed tan, friable bladder neck mass. After transurethral resection, pathology identified a solitary fibrous tumor with diffuse mitotic features concerning for malignancy. Due to these features and the site of tumor being the trigone and bladder neck, a partial cystectomy was unfeasible. The patient elected to undergo a radical cystoprostatectomy with ileal conduit diversion. A 6.5 X 4 X 4.5 cm butterfly shaped tan-white solid mass at the junction of the bladder neck was present without invasion into the prostate (Figure 2).

Histologically, this was a spindle cell neoplasm composed of hypercellular sheets of atypical cells with transition areas of less cellularity. The hypercellular areas had cells with enlarged, hyperchromatic, nuclei with minimal cytoplasm and increased mitoses (up to 13 per 10 high power fields) (Figure 3a). The less cellular areas were characterized by scattered, thin-walled,

Figure 1: CT with contrast demonstrating bladder neck mass and enlarged prostate

ectatic vessels with irregular branching and spindled-to-oval cells with bland nuclei, scarce mitoses and variable amounts of sclerotic collagen (Figure 3b). By immunohistochemistry, the neoplastic cells were positive for STAT6 a marker highly sensitive and specific for the NAB2-STAT6 gene fusion found in SFTs. Additionally, the tumor was positive for BCL2 and CD34 and negative for S100, cytokeratin AE1/AE3, desmin, and actin. Due to the elevated mitotic rate (>4 mitoses per 10 hpf), hypercellularity, and cell atypia, a diagnosis of a malignant HPC/SFT was rendered. The margins were free of tumor. No adjuvant chemotherapy was administered.

Discussion

Hemangiopericytoma/Solitary Fibrous Tumor is an exceedingly rare neoplasm of mesenchymal origin that accounts for 2.5 % of all soft tissue tumors [3,4]. Previous literature shows that hemangiopericytomas share various histologic and immunohistochemical features with solitary fibrous tumors [9]. This commonality along with a shared fibroblastic origin prompted a reclassification to HPC/SFT in the World Health Organization meeting in 2002 [2]. These masses have a pseudocapsule and a staghorn vascular network surrounded by spindle cells [10]. The final diagnosis necessitates the presence of CD34, a monoclonal

antibody against the human hematopoietic progenitor cell antigen [11]. Clinically, HPC/SFTs present as a slow growing mass in middle aged adults of both sexes. The retroperitoneum, pleura, deep soft tissues of proximal extremities, abdominal cavity, trunk, and head and neck are the most common sites [9]. The abdominal cavity and retroperitoneum are the two most common sites for malignant HPC/SFTs [3,11] with the lungs, liver, and bones being the primary metastatic sites [12]. Bladder HPC/SFTs are rare; therefore we investigated the available literature for non-bladder HPC/SFTs to identify any trends in management or recurrence. Due to their solitary, indolent, and locally infiltrative nature, radical resection appears to be the treatment of choice in the CNS, head and neck, abdomen and other sites with chemotherapy and radiation having limited roles [12,13,14].

Two malignant bladder HPC/SFT cases have been reported out of 10 total cases in the literature. The median age of onset for bladder HPC is 50 years old with age ranging from 29 to 72 at the time of diagnosis [15]. HPC in other parts of the body seems to have an equal predisposition towards men and women which is similar in our literature review finding (6 female and 4 male). Three patients were diagnosed following episodes of gross hematuria, although five patients described lower urinary tract symptoms including retention, dysuria, and frequency (Table 1). Others presented with symptoms unrelated to the genitourinary tract.

The workup for HPC does not differ from the standard hematuria workup including a cystoscopy and axial imaging of the abdomen and pelvis. The treatment and followup for bladder HPC are uncertain owing to the rarity of the disease. Surgical resection has been as the mainstay of treatment. Other management options included radiation, transurethral resection, partial and radical cystectomy [9]. One transurethral resection and nine cystectomies have been performed previously (Table 1). Transurethral resections may not provide definitive therapy or excise all of the tumor likely due to the non-epithelial origin of the tumor; the sole case also demonstrates recurrence at 2 years [15,16]. We believe en bloc resection may provide patients with more durable oncologic outcomes. Adjuvant therapy should be considered in HPCs demonstrating malignant features, although the best modality is yet to be determined [17]. Chemotherapy has been tried in one case with metastasis, but no benefit was demonstrated [17]. Adjuvant radiotherapy appeared to be successful in the case of a 45 year old status post partial cystectomy [18]. We also investigated other genitourinary HPC/SFTs to guide us regarding adjuvant therapy. In Renal HPC/SFTs, neither chemotherapy nor radiotherapy has been effective adjuvant modalities [19,20]. Given the limited data available in adjuvant therapy, we recommend considering it in a case by case manner in malignant HPC/SFTs.

Long-term follow-up is not available from all previous studies making prognosis uncertain. Outcomes in these studies also appear varied. The longest documented survival without evidence of recurrence is 12 years [21] in a 54 year old man who underwent partial cystectomy for benign HPC/SFT. However, lung metastasis has been reported as far as 9 years

Figure 2: Posterior view of bladder mass (open).

Figure 3: A) Malignant solitary fibrous tumor with densely cellular sheets of atypical, hyperchromatic cells and increased mitotic figures, 400x magnification (a). Areas having typical features of solitary fibrous tumor including thin-walled, branched ectatic vessels and bland-appearing spindled-to-oval cells, 100x magnification (b).

Table 1: Characteristics and treatments of patients with HPC/SFT, n=11 including current case. 1 case treatment unknown.

Gender (Male, Female)	Presenting Symptoms	Treatment	Outcome
Female	Anemia and weight loss	Complete cystectomy	No recurrence at 2 years
Female	Hypoglycemia	Partial cystectomy with palliative radiation for metastases	Metastases at 7 years
Female	Right lower quadrant abdominal pain	Partial cystectomy	No recurrence at 6 months
Female	Acute urinary retention	Transurethral resection	Recurrence at 2 years
Female	Painless hematuria, frequency, and suprapubic discomfort	Partial cystectomy	Passed at 3 days, secondary to a pulmonary embolus from tumor thrombus
Female	No symptoms	Partial cystectomy	No recurrence at 2 years
Male	Left groin pain and urinary frequency	Partial cystectomy and adjuvant radiotherapy	No recurrence at 2 years
Male	Right groin mass with pain. Urinary frequency and dysuria.	Partial cystectomy and chemotherapy for metastases	Metastases at 9 years
Male	Right lower quadrant abdominal mass, painless hematuria	Partial cystectomy	No recurrence at 12 years
Male*	Painless hematuria and urinary retention	Complete cystectomy	No recurrence at 1 year
NA	NA	NA	NA

* represents current case

following initial surgery [22]. Therefore, lifelong surveillance is recommended in patients with HPC/SFT of the urinary bladder. Limited experience in bladder HPC/SFTs makes treatment and management uncertain. We present a review of published literature with management recommendation based on previous treatment pathways. Due to the reports of local recurrence with transurethral resections and non-epithelial origin of the tumor, we believe complete resection with partial or complete cystectomy is the choice for best curative rate. For adjuvant therapy, the effectiveness of chemotherapy and radiotherapy remain debatable secondary to the small number of cases.

Conflict of Interest

There are no conflicts of interests to declare on behalf of any of the authors. Informed Consent was obtained from the patient mentioned in the case report.

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