Bladder Schwannoma - A Case Presentation

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ABSTRACT

Bladder schwannomas are exceedingly rare, benign or malignant, nerve sheath tumors that are most often discovered in patients with a known diagnosis of Neurofibromatosis type 1 (NF1). A few sporadic case reports of bladder schwannoma have been published in urologic, obstetric/gynecologic, and pathologic journals. However, this is the first case report in the radiologic literature where computed tomography imaging and radiology-specific descriptions are discussed. Furthermore, the patient presented in this case is only the fifth published patient without NF1 to be diagnosed with a bladder schwannoma, to the best of our knowledge.

CASE REPORT

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The patient is a 31 year old male with a history of recurring nephrolithiasis who had presented to the emergency department with colicky flank pain and trace hematuria on several occasions over a three year period. Non-contrast computed tomography (CT) scans of the abdomen and pelvis performed on the patient's first two visits to the emergency department demonstrated non-obstructive nephroliths bilaterally with no other intra-abdominal or intra-pelvic pathology (Figure 1). On both occasions, the patient was treated symptomatically and discharged from the emergency department.

Fifteen months after his second visit to the emergency department, the patient presented with hematuria, right flank pain, and right upper quadrant pain. Given his history of nephrolithiasis, a non-contrast CT of the abdomen and pelvis was performed. The CT demonstrated similar appearing bilateral non-obstructing nephroliths and a homogeneous, well-circumscribed mass with a density of 32 Hounsfield units (HU) on the left anterolateral bladder wall (Figure 2). The mass was relatively isodense to the bladder wall and measured 1.7 X 1.1 centimeters in axial dimensions. Assessment of bladder wall thickening was mildly limited secondary to a lack of intravenous or intravesicular contrast, but no wall thickening was seen adjacent to the lesion. No associated inflammatory fat stranding was present to suggest extra-

Radiology Case. 2012 Dec; 6(12):26-31

vesicular extension and no suspicious lymphadenopathy was present. The differential included a bladder neoplasm and blood clot.

After the bladder mass was diagnosed, the patient was treated symptomatically for his pain and the urology service was consulted. The urology service believed the lesion likely represented a neoplasm given that the patient had minimal risk factors for developing a blood clot. No further cross-sectional imaging was requested by the urology service prior to proceeding with cystoscopy. Three days later, flexible cystoscopy confirmed there was a pedunculated left anterolateral bladder wall mass, which was presumed to represent bladder carcinoma. Biopsies of the lesion were sent to pathology for further evaluation (Figure 3).

On macroscopic examination, the submucosal mass appeared tan, smooth and rubbery. The mass was sectioned and stained with Hematoxylin and Eosin (H&E) for further evaluation. Light microscopy revealed a spindle cell neoplasm with areas of hypocellularity (Antoni B) and areas of dense cellularity (Antoni A). Within the densely cellular areas, palisading nuclei alternated with pink, nuclear free zones (Verocay bodies) (Figure 4). These findings are highly characteristic of a schwannoma. Although S100 immunohistochemistry is pathognomonic for schwannoma, S100 immunohistochemistry was not performed on the biopsy sample, as the appearance on H&E stained slides was consistent with a schwannoma and the pathologist did not feel additional processing with immunohistochemistry would improve the diagnostic yield.

The schwannoma was later surgically excised. The patient had no immediate post-operative complications. He was followed by the urology service for 8 months without tumor recurrence before moving out of state.

DISCUSSION

Although schwannomas are more often located in the head and neck (40-50%), they can occur anywhere in the body where a nerve sheath is present, including the bladder [1]. Although transitional cell carcinoma (>90%), squamous cell carcinoma (5%), and adenocarcinoma (2%) account for the majority of bladder wall masses [1, 2], other less common bladder neoplasms to be considered in the differential include neurofibromas, leiomyomas, solitary fibrous tumors, and schwannomas.

Primary schwannomas of the bladder are exceedingly rare tumors, representing <0.1% of all bladder tumors [3]. Bladder Schwannomas have no gender predilection, are most common in the 4th-6th decade, and are often associated with Neurofibromatosis type 1 (NF1) [3, 4, 5]. These tumors are usually slow growing and benign, although malignant variants have been reported (< 5%) [6]. Positive S100 immunohistochemistry is the pathognomonic pathology finding of a schwannoma [7]. Two highly characteristic patterns for schwannoma can be seen on H&E staining. Antoni A areas consist of compact intersecting spindle cells with elongated nuclei arranged in parallel bundles, incomplete whorls and complete whorls (Verocay bodies) [8]. Antoni B areas are composed of loosely arranged spindle cells [8].

Journal of Radiology Case Reports

The CT appearance of a schwannoma is non-specific. However, a few characteristic features exist. Bladder schwannomas appear isodense or hypodense to surrounding muscle [9]. Schwannomas are not typically associated with calcifications [5]. Their enhancement tends to be dense and homogeneous when small and increasingly heterogeneous as they enlarge [1]. However, enhancement patterns have not been shown to be a reliable way to differentiate schwannomas from other bladder tumors [10]. Despite the inability to adequately differentiate various bladder tumors, contrastenhanced CT is excellent in detecting up to 97% of all bladder neoplasms [11].

Magnetic resonance imaging (MRI) is slightly more sensitive than CT for the evaluation of suspected schwannomas, but differentiation between a bladder schwannoma and carcinoma remains difficult. Both schwannomas and carcinomas are usually isointense to skeletal muscle on T1 weighted imaging (T1WI) and isointense to slightly hyperintense to skeletal muscle on T2 weighted imaging (T2WI) [2, 9]. Given their similar appearance on both CT and MRI, clinical history is of the upmost importance. When a bladder mass with characteristics similar to bladder carcinoma is seen in a patient with NF1, it would reasonable to include bladder schwannoma in the differential, given that a schwannoma can appear similar to bladder carcinoma and NF1 is the most common risk factor for developing a bladder schwannoma.

The two other entities to consider in the differential for a bladder mass are an intravesicular blood clot and extrinsic mass. A blood clot typically will have a density between 30-50 HU, which may be slightly hypodense, isodense or slightly hyperdense when compared to surrounding skeletal muscle, which usually has a density of 40 HU. Lack of contrast enhancement and resolution over time are the two distinguishing features of a blood clot, which help differentiate it from a bladder wall neoplasm. An extrinsic mass causing urinary outflow obstruction can result in bladder wall trabeculation, which can mimic a neoplasm. Contrast enhanced imaging will usually demonstrate a symmetrically thickened bladder wall without a discrete enhancing mass.

The initial differential on the non-contrast CT included a bladder neoplasm and intravesicular clot. Urinary outflow obstruction was not considered in the differential because the lesion appeared discrete, the patient's prostate was normal in size, and there was no associated bladder wall thickening. After the patient underwent cystoscopy, the presumed diagnosis was bladder carcinoma. Interestingly, a bladder schwannoma was diagnosed in this patient with no prior history or clinical features suggestive of NF1. The patient underwent surgical resection, which is the treatment of choice for bladder schwannomas as they may lead to ureteral obstruction and have a small chance of malignant degeneration (<5%) [1, 12].

TEACHING POINT

Bladder schwannomas tend to have a similar appearance to bladder carcinomas on both CT and MRI. Given that bladder schwannomas are so rare, they usually should not be included in the differential. However, in a patient with known neurofibromatosis type 1, it would be reasonable to include bladder schwannoma in the differential given that neurofibromatosis type 1 is the most common risk factor associated with bladder schwannoma. Since bladder carcinomas are so much more common, they will be the most common bladder tumors diagnosed in patients with neurofibromatosis type 1.

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FIGURES

Figure 1. 31 year old male diagnosed with a bladder schwannoma. Select axial images from non-contrast abdominal CTs (soft tissue windows) performed on the patient's first (left) and second (right) visit to the emergency department. The first visit was approximately 3 years prior to his diagnosis of bladder schwannoma, and the second visit was 15 months prior to his diagnosis. No bladder mass is demonstrated on either CT. (Protocol: 16 slice, 120 KVP, 119 mA, 5 mm slice thickness, non-contrast).

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Figure 2. 31 year old male diagnosed with a bladder schwannoma. Select axial image from a non-contrast abdominal CT in soft tissue windows demonstrates a homogeneous, well-circumscribed, solid mass on the left anterolateral bladder wall (right). A magnified view of the same lesion is also displayed (left). The mass is isodense to the bladder wall and measures 1.7 X 1.1 cm in axial dimension. (Protocol: 16 slice, 120 KVP, 123 mA, 5 mm slice thickness, non-contrast).



Figure 3 (left). 31 year old male diagnosed with a bladder schwannoma. Single image obtained during a cystoscopy demonstrates a tan, smooth-appearing, submucosal mass, which has been partially removed during a biopsy procedure.



Figure 4. 31 year old male diagnosed with a bladder schwannoma. Low (left) and High (right) magnification hematoxylin and eosin stains from the bladder mass biopsy. Low magnification demonstrates a spindle cell neoplasm with densely cellular areas (Antoni A), loose hypocellular areas (Antoni B), and calcifications. High magnification demonstrates palisading of spindle cell nuclei alternating with nuclear free zones (Verocay bodies) within the Antoni A areas. These pathologic findings are highly characteristic of a schwannoma.

Etiology	Unknown	
Incidence	Extremely rare (<0.1% of bladder wall tumors)	
Gender Ratio	No predilection	
Age	4th-6th decade	
Risk Factors	Most are associated with neurofibromatosis type 1	
Treatment	Surgical excision	
Prognosis	Favorable if excised and no malignant degeneration is found pathologically	
Imaging Findings	•CT shows a solitary, non-invasive (if benign), lobulated mass, isodense to the bladder wall, and	
	usually without calcifications.	
	•Small schwannomas tend to enhance homogeneously, while larger schwannomas are more	
	heterogeneous.	

Table 1: Summary table: This table summarizes the key aspects of and imaging findings associated with a bladder schwannoma (3, 5, 12).

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Differential	СТ	MR
Bladder Schwannoma	Solitary, non-invasive (if benign), lobulated mass,	T1WI isointense, T2WI isointense to slightly
	isodense to the bladder wall, and usually without	hyperintense to skeletal muscle. Small
	calcifications. Small schwannomas tend to	schwannomas tend to enhance
	enhance homogeneously, while larger	homogeneously, while larger schwannomas
	schwannomas are more heterogeneous.	are more heterogeneous.
Bladder Carcinoma	Soft tissue mass with mild/moderate delayed (60-	T1WI isointense, T2WI isointense to slightly
(adenocarcinoma or	80 s) enhancement, +/- calcifications, +/-	hyperintense to skeletal muscle. Enhancement
squamous cell	perivesical invasion.	patterns are variable and similar to
carcinoma)		schwannomas.
Blood Clot	Often mobile, non-enhancing mass with variable	Heterogeneous, non-enhancing mass.
	density depending on age of blood products. Clot	
	resolves with time.	
Extrinsic mass (Benign	Mass effect causes bladder wall trabeculation	Mass effect causes bladder wall trabeculation
prostatic hypertrophy)	without discrete enhancing mucosal mass.	without discrete enhancing mucosal mass.

Table 2: Differential Table: Differential diagnosis table for bladder schwannoma. The differential for a bladder schwannoma includes bladder wall carcinoma, blood clot, and extrinsic masses such as benign prostatic hypertrophy (1, 3, 7, 11).

ABBREVIATIONS

CT: Computed tomography H&E: Hematoxylin and Eosin HU: Hounsfield units MRI: Magnetic resonance imaging NF1: Neurofibromatosis type 1 T1WI: T1 weighted imaging T2WI: T2 weighted imaging

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KEYWORDS

Bladder mass; bladder schwannoma; neurofibromatosis; schwannoma

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