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Case study

Benign perivascular myoid cell tumor (myopericytoma) of the urinary tract: a report of 2 cases with an emphasis on differential diagnosis

Ming Zhao MD[a], Sean R. Williamson MD[b], Ke Sun MD[c], Yin Zhu MD[a], Changshui Li MD[a], Wenping Xia MD[d], Honggang Qi MD[e], Lisha Wang MD[f], Konstantinos Linos MD[g], Liang Cheng MD[g,*]

[a]*Department of Pathology, Ningbo Yinzhou Second Hospital, Ningbo, Zhejiang 315100, China*
[b]*Department of Pathology, Henry Ford Health System, Detroit, MI 48202, USA*
[c]*Department of Pathology, the First Affiliated Hospital, Zhejiang University College of Medicine, Hangzhou, Zhejiang 310003, China*
[d]*Department of Radiology, Ningbo Yinzhou Second Hospital, Ningbo, Zhejiang 315100, China*
[e]*Department of Urology, Ningbo Yinzhou Second Hospital, Ningbo, Zhejiang 315100, China*
[f]*Department of Pathology, Fudan University Shanghai Cancer Center, Shanghai 200032, China*
[g]*Department of Pathology and Laboratory Medicine, Indiana University School of Medicine, Indianapolis, IN 46202, USA*

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Summary Myopericytoma is a benign mesenchymal neoplasm thought to comprise part of a spectrum of perivascular myoid cell neoplasms with myofibroma, angioleiomyoma, and glomus tumor. We describe 2 such neoplasms involving the urinary tract: 1 incidentally identified in the kidney of a 59-year-old woman and 1 in the urinary bladder of a 52-year-old woman who presented with urinary frequency and dysuria. Histologically, the bladder tumor was composed of numerous blood vessels surrounded by plump perivascular myoid cells, as in subcutaneous myopericytoma. The renal tumor showed similar morphology centrally and a symplastic glomus tumor–like growth pattern at the periphery. Immunohistochemically, both tumors were reactive for markers of smooth muscle differentiation, such as smooth muscle actin and caldesmon/calponin but negative for CD34, cathepsin K, and S100 protein. Both patients are free of disease 14 and 39 months after resection, respectively. Our findings broaden the morphologic spectrum of myopericytoma.

1. Introduction

Myopericytoma is a benign neoplasm composed of a perivascular arrangement of cytologically uniform oval- or spindle-shaped cells with a myoid appearance and immunophenotype. These tumors are thought to comprise part of a morphologic spectrum of perivascular myoid cell neoplasms that also includes myofibroma/myofibromatosis (infantile hemangiopericytoma), angioleiomyoma, and glomus tumor [1-7]. In the past, myopericytomas have likely been classified as solitary myofibroma or hemangiopericytoma; however, it is now recognized that the term *hemangiopericytoma* characterizes a morphologic growth pattern shared by multiple

* Corresponding author. Department of Pathology and Laboratory Medicine, Indiana University School of Medicine, IUHPL-4044, Indianapolis, IN 46202, USA.
E-mail address: liang_cheng@yahoo.com (L. Cheng).

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subtypes of mesenchymal tumors rather than a distinctive clinicopathological entity. Thus, this terminology has been largely removed from the most recent classification of tumors of soft tissue by the World Health Organization [1]. In this updated classification system, myopericytoma and myofibroma are grouped together under the same heading, and the latter appears as a subtype of the former.

Myopericytomas are typically found in the skin and superficial soft tissues, most commonly in the extremities or occasionally in the head and neck or trunk, forming a solitary, painless, and slow-growing mass [3,4,6,8]. Rarely, these tumors have been reported to occur in other sites, including the oral cavity, nasal cavity, thorax and lung, heart, gastrointestinal tract, and brain [5]. In the urinary tract, only 2 examples of renal myopericytoma have been reported in the English literature to date [9,10]. Herein, we describe 2 such perivascular myoid cell tumors arising in the urinary tract: 1 in the kidney, with an unusual morphologic appearance eliciting a broad spectrum of differential diagnoses and 1 in the urinary bladder, to our knowledge the first report of urinary bladder myopericytoma.

2. Case report

2.1. Case 1

A 59-year-old woman was incidentally found to have a left-sided renal mass during ultrasonographic workup for cholecystitis and cholelithiasis. Her medical history included hypertension of 10 year-duration. Subsequent computed tomography demonstrated a well-demarcated, heterogeneously enhancing exophytic mass with a maximum diameter of 3.5 cm in the left midkidney (Fig. 1A). The mass bulged from the renal contour but did not invade perinephric fat, the renal vein, or the inferior vena cava. Imaging studies revealed no evidence of metastatic disease. Given the suspicion for renal cell carcinoma, the patient underwent laparoscopic left radical nephrectomy with no postoperative complications, and she was discharged 2 weeks after the surgery. After resection of the mass, no change in her hypertension was noted. No residual, recurrent, or metastatic tumor was identified at most recent follow-up of 14 months.

2.2. Case 2

A 52-year-old woman presented with urinary frequency and dysuria of 4 year-duration, worsening over a period of 1 month, without gross hematuria or weight loss. Medical history was unremarkable, including negative testing for human immunodeficiency virus. Abdominal computed tomography revealed a well-circumscribed, heterogeneously enhancing mass in the posterior wall of the urinary bladder. Imaging did not reveal local invasion or regional or distant metastasis. Urine cytology was negative for malignant cells. Partial cystectomy was performed, and the patient recovered uneventfully. No residual, recurrent, or metastatic tumor was identified at most recent follow-up of 39 months.

Table Results of immunohistochemical staining and in situ hybridization analysis

Antibody (source)	Case 1 (kidney)	Case 2 (urinary bladder)
SMA (Dako, Carpinteria, CA)	Diffusely, strongly positive	Diffusely, strongly positive
Desmin (Dako)	Patchy, weakly positive	Patchy, weakly positive
Caldesmon (Dako)	Diffusely, strongly positive	Diffusely, strongly positive
Vimentin (Dako)	Diffusely, strongly positive	Diffusely, strongly positive
Calponin (Dako)	Diffusely, strongly positive	Diffusely, strongly positive
Cathepsin K (Abcam, Cambridge, MA)	Negative	Negative
CD34 (Dako)	Negative	Negative
CD31 (Dako)	Negative	Negative
AE1/3 (Dako)	Negative	Negative
EMA (Dako)	Negative	Negative
C-kit (Dako)	Negative	Negative
NSE (Dako)	Diffusely, weakly positive	Negative
S100 protein (Dako)	Negative	Negative
HMB45 (Dako)	Negative	Negative
Melan-A (Dako)	Negative	Negative
Chromogranin A (Dako)	Negative	Negative
Synaptophysin (Dako)	Negative	Negative
CD10 (Dako)	Negative	Negative
Estrogen receptor (Dako)	Negative	Diffusely, strongly positive
Progesterone receptor (Dako)	Negative	Diffusely, strongly positive
EBER-ISH (Novocastra, Newcastle, UK)	Negative	Negative

Abbreviations: SMA, smooth muscle actin; NSE, neuron-specific enolase; EBER-ISH, in situ hybridization for Epstein-Barr virus–encoded RNA.