

## Oncology

## Pelvic hemangiopericytoma: A case report and review of the literature

Anirban P. Mitra<sup>a,b,\*</sup>, Marissa Maas<sup>a</sup>, Daniel E. Zainfeld<sup>a,b</sup>, Anne K. Schuckman<sup>a,b</sup><sup>a</sup> Institute of Urology, Keck School of Medicine, University of Southern California, Los Angeles, CA, USA<sup>b</sup> Department of Urology, Los Angeles County + University of Southern California Medical Center, Los Angeles, CA, USA

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## ABSTRACT

Extrauterine pelvic hemangiopericytomas are rare tumors with malignant potential. Herein, we report a case of a 55-year-old male who presented with hydronephrosis and obstructive urinary symptoms. Cystoscopy, imaging and biopsy were unable to provide a definitive preoperative diagnosis. He underwent pelvic mass resection, and final pathology revealed malignant hemangiopericytoma. The patient is being closely monitored with serial imaging, and remains disease-free at 23 months of post-operative follow-up without adjuvant therapy. Our experience and evidence from the existing literature suggests that given the rarity of these tumors, adherence to standard oncologic principles is necessary to ensure adequate resection and appropriate follow-up.

## Introduction

Hemangiopericytomas are rare vascular neoplasms related to smooth muscle perivascular cells known as pericytes. Initially described by its histopathologic, albeit nonspecific, staghorn vascular pattern, the diagnosis included several unrelated benign and malignant entities. With the advent of immunohistochemistry, histologic mimics were excluded and hemangiopericytoma became a distinct entity, although largely a diagnosis of exclusion. Grading is based on the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) system that accounts for tumor differentiation, necrosis, and mitotic activity. Modern reports of extrauterine pelvic hemangiopericytomas are extremely rare. We report a case of a male patient who presented with obstructive urinary symptoms and hydronephrosis due to a pelvic hemangiopericytoma. We also summarize published reports of extrauterine pelvic hemangiopericytomas to date to raise awareness regarding the diversity of symptoms and diagnostic findings associated with this disease.

## Case report

A 55-year-old African American male with history of hypertension was referred for lower urinary tract symptoms (LUTS), new-onset left hydronephrosis and worsening renal function. His estimated glomerular filtration rate (eGFR) worsened from 84 mL/min to 52 mL/min within one year. At initial presentation, he reported nocturia, weak stream, severe urgency, and urge incontinence. Surgical history included take down of a left lower quadrant temporary colostomy that was initially

created after emergent surgery for an abdominal gunshot wound 20 years ago. He reported being an active smoker and maternal history of kidney cancer. Initial renal ultrasound ordered by his primary care provider reported left hydronephrosis and possibly an “enlarged prostate”. Prostate-specific antigen measured 1.16 ng/mL. Uroflowmetry showed Qmax of 16.9 mL/sec and postvoid residual of 293 mL. Cystoscopy revealed distorted bladder lumen anatomy without a discernible interureteric ridge, and the left lateral wall showing evidence of extraluminal compression. The left ureteral orifice could not be identified.

CT imaging showed a centrally necrotic 9.3 cm × 9.7 cm × 10.3 cm pelvic mass posterior to the urinary bladder and superior to the prostate exerting a mass effect, and compressing the left ureter with resultant severe hydronephrosis with cortical thinning (Fig. 1A and B). No radiologic evidence of metastatic disease was seen in the chest, abdomen or pelvis. Percutaneous biopsy of the mass showed rare atypical spindle cells that were immunoreactive for CD34, FLI-1 and CD99, suggesting an epithelioid mesenchymal neoplasm. A left nephrostomy tube was placed to salvage renal function. MRI demonstrated a necrotic pelvic mass displacing the bladder to the right and abutting the left seminal vesicle (Fig. 1C and D).

The patient underwent open laparotomy for mass resection under general anesthesia. In supine position, a lower midline incision was made and the retroperitoneal space was entered. Ureters were identified bilaterally and mobilized to the pelvis. The peritoneum was incised and the mass was mobilized off the bladder; it was not adherent to or invading the bladder. The right ureter was traced up to its insertion into

\* Corresponding author. Institute of Urology, University of Southern California, 1441 Eastlake Avenue, Suite 7416, MC 9178, Los Angeles, CA, 90033, USA.  
E-mail address: [apmitra@gmail.com](mailto:apmitra@gmail.com) (A.P. Mitra).

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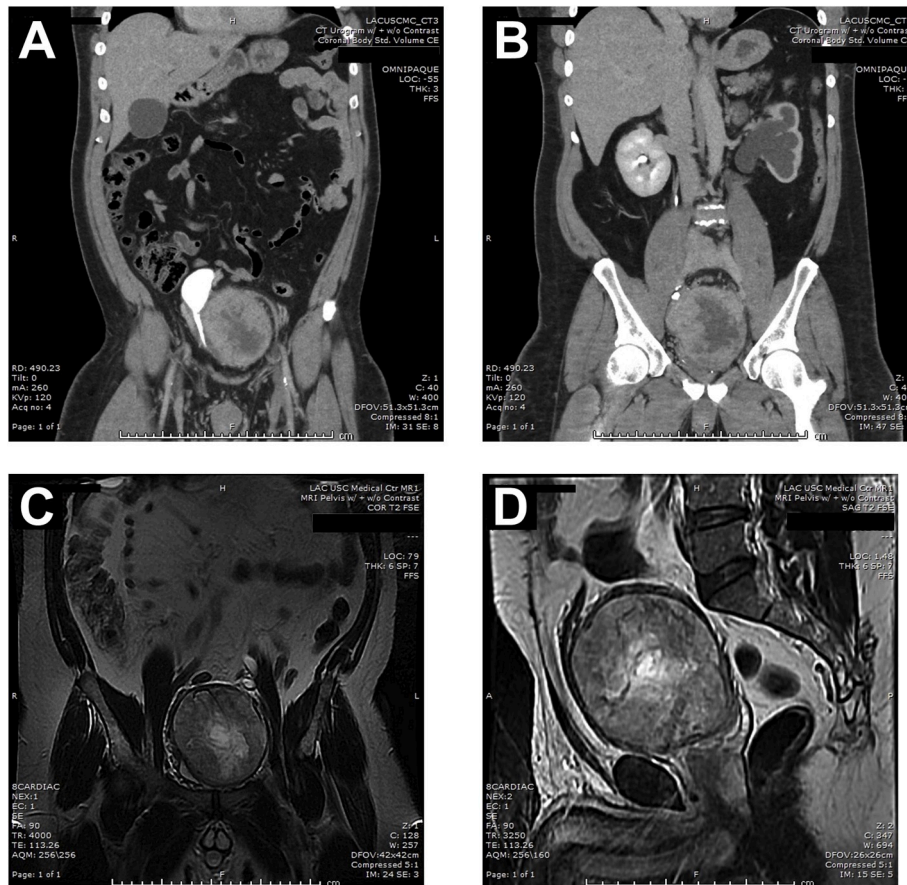
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**Fig. 1.** Excretory phase computed tomography showed (A) a centrally necrotic pelvic mass displacing a contrast-filled urinary bladder to the right and (B) resulting in severe left hydronephrosis with cortical thinning. (C) Coronal and (D) sagittal views on magnetic resonance imaging confirmed presence of a pelvic mass anterior to the rectum and displacing the urinary bladder.

**Table 1**  
Published reports of extrauterine pelvic hemangiopericytomas.

Age (years)/Sex	Symptoms	Size (cm)	Location/Primary Treatment, Any Salvage Treatment	Follow-up (months)/Last Clinical Status	Report/Year/PMID
56 M	Mass	4	Adherent to rectum <sup>1</sup>	N/A	Bacon et al., 1950 (15429830)
34 F	Rectal bleed	3.5 × 2.5	Sigmoid, mesenteric border <sup>1,5</sup>	60 <sup>b</sup>	Ault et al., 1951 (14866696)
40 F	Pain	5	Cul de sac <sup>1,5</sup>	11 <sup>b</sup>	Forman et al., 1952 (14914815)
43 M	Mass	20 × 11 × 10.5	Pelvis <sup>1,5,6</sup>	78 <sup>c</sup>	Wise et al., 1952 (14932614)
47 F	Pain	8.5 × 7.8 × 6.7	Left pelvis <sup>1,6</sup>	N/A	Stout et al., 1956 (13296384)
11 M	Mass	N/A	Left iliac fossa <sup>2</sup>	119 <sup>c</sup>	Stout et al., 1956 (13296384)
50 F	Mass	4.5 × 3 × 3.5	Adherent to rectum <sup>1</sup>	18 <sup>a</sup>	Slattery et al., 1956 (13313911)
16 F	Hypoglycemia	N/A	Left pelvis <sup>1</sup>	11 <sup>c</sup>	Howard et al., 1959 (13630072)
50 F	N/A	4.5	Adherent to rectum <sup>1</sup>	48 <sup>a</sup>	Fisher et al., 1960 (13699977)
39 F	Pain	15 × 10 × 10	Retroperitoneal <sup>1,5</sup>	39 <sup>a</sup>	Spiro et al., 1964 (14207337)
73 F	None	25 × 15 × 10	Retroperitoneal <sup>4</sup>	96 <sup>a</sup>	Wilbanks et al., 1975 (1200042)
58 M	Bladder outlet obstruction	N/A	Pelvis <sup>1</sup>	N/A <sup>a</sup>	Roberts et al., 1977 (883072)
29 F	Dyspareunia, dysuria	N/A	Retroperitoneal <sup>1</sup>	N/A	Kaude et al., 1980 (6448795)
57 M	Pain	8	Pelvis <sup>1</sup>	N/A	Yoo et al., 1986 (3943360)
59 F	Pain	15 × 15	Pelvis <sup>2,6</sup>	24 <sup>a</sup>	Kehagias et al., 1999 (9933402)
40 M	None	14 × 9	Pelvis <sup>1</sup>	N/A	Rosenblatt et al., 2001 (11586234)
19 M	Pain	7 × 6 × 9	Right pelvic wall <sup>3,5</sup>	88 <sup>a</sup>	Unal et al., 2002 (11816000)
42 F	Pain	7 × 7 × 5	Inguinal <sup>3,5</sup>	64 <sup>a</sup>	Unal et al., 2002 (11816000)
34 M	Pain, mass	10 × 12 × 10	Pelvic wall <sup>3,5</sup>	13 <sup>a</sup>	Unal et al., 2002 (11816000)
58 F	Pain, constipation, frequency	N/A	Right ischiorectal fossa, ischiopubic ramus <sup>2</sup>	12 <sup>a</sup>	Perdikakis et al., 2011 (22933930)
63 F	Pain	12 × 9 × 9.4	Pouch of Douglas <sup>1</sup>	N/A <sup>a</sup>	Álvarez Abad et al., 2017 (26912344)
55 M	Bladder outlet obstruction	9.3 × 9.7 × 10.3	Pelvis <sup>1</sup>	23 <sup>a</sup>	Mitra et al., 2020 (Present study)

**Abbreviations:** F, female; M, male; N/A, not available; PMID, PubMed identifier, available at [pubmed.ncbi.nlm.nih.gov](https://pubmed.ncbi.nlm.nih.gov).

<sup>1</sup>Surgical resection.

<sup>2</sup>Radiotherapy.

<sup>3</sup>Hyperthermic pelvic chemotherapy perfusion.

<sup>4</sup>N/A.

<sup>5</sup>Salvage resection.

<sup>6</sup>Adjuvant or salvage radiotherapy.

<sup>a</sup>Alive with no evidence of disease.

<sup>b</sup>Alive with disease.

<sup>c</sup>Dead of disease.

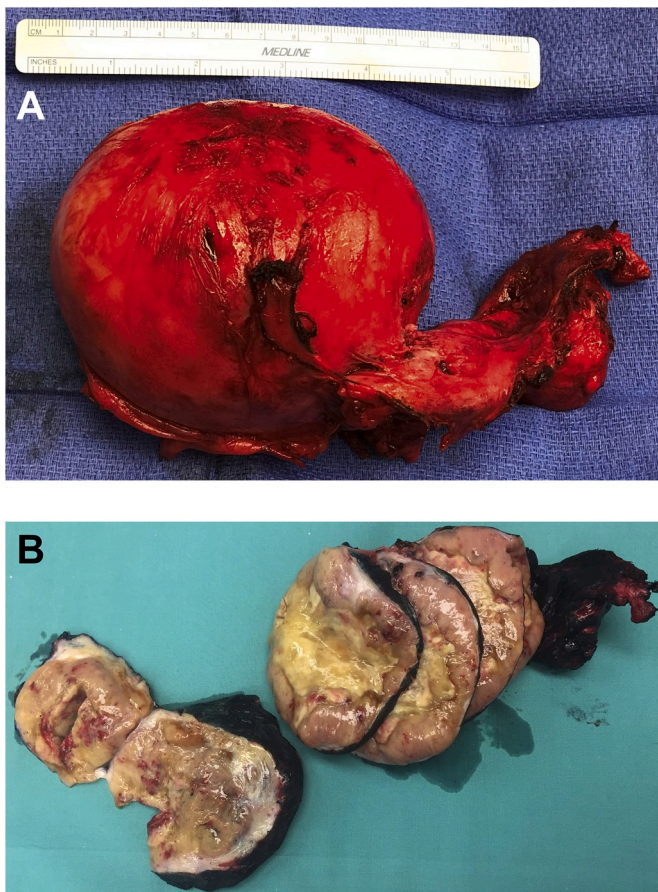
the bladder. However, the distal left ureter was densely adherent to the mass; it was therefore divided proximally and removed with the mass *en bloc*. The left ureter was then extravesically reimplanted to the bladder dome in a tension-free manner. The mass did not appear to involve or invade any other pelvic structures. Bilateral ureteral stents were placed. A urethral catheter was placed in the bladder, and a Blake drain was placed in the retropubic space. Operative time was 365 mins with estimated blood loss of 600 mL. Patient had slow return of bowel function but otherwise uneventful hospital course and was discharged on post-operative day (POD) 4 after removal of the nephrostomy tube. Urethral catheter and Blake drain were removed after cystogram confirmed no contrast extravasation on POD 18; ureteral stents were removed on POD 32. He reported significant improvement of LUTS after recovering from surgery.

Gross evaluation of the 490 gm specimen revealed a well-circumscribed fleshy mass with a central necrotic lesion (Fig. 2). Final histopathological diagnosis was malignant hemangiopericytoma, FNCLCC grade 2. Specimen showed immunoreactivity for vimentin and CD34, with 15% cells being Ki67-positive. Repeat imaging at four months showed improvement of left hydronephrosis with eGFR of 64 mL/min. The patient remains on surveillance with serial CT imaging of the chest, abdomen and pelvis every 3–4 months. He remains disease-free at 23 months of post-operative follow-up. No adjuvant chemotherapy or radiation has been administered.

## Discussion

Solitary fibrous tumors comprise a histologic spectrum of mesenchymal neoplasms that are found in many locations, but are most commonly seen in the pleura, peritoneum, meninges and lower extremities.<sup>1</sup> Hemangiopericytoma, a rare subset of malignant solitary fibrous tumors, are soft-tissue sarcomas that originate from mesenchymal cells with pericytic differentiation. Pericytes are arranged around capillaries and postcapillary venules; consequently, hemangiopericytomas may occur anywhere capillaries are found. These are usually slow-growing and painless. Extrauterine pelvic hemangiopericytomas are extremely rare; our literature review suggests that common presenting symptoms include vague abdominopelvic pain and those associated with mass effect (Table 1).

Hemangiopericytomas appear to be equally distributed among genders and age groups. Imaging is usually non-specific and may show a homogenous mass surrounded by a capsule with vascular enhancement. Immunohistochemical findings include reactivity for CD34, CD99 and vimentin.<sup>2,3</sup> Clinical behavior may range from a benign course to malignant transformation. Since most hemangiopericytomas are not metastatic at presentation, surgery remains the gold standard treatment and is associated with high disease-free survival rates.<sup>4</sup> However, the unpredictable tumor behavior necessitates close follow-up and adjuvant therapy for aggressive disease. Adjuvant radiation may confer a survival benefit, especially in patients with high-grade disease.<sup>5</sup>



**Fig. 2.** (A) Grossly, the 10.5 cm pelvic mass appeared well-circumscribed with a smooth surface. (B) Sectioning revealed a fleshy mass with a central necrotic lesion.

### Conclusion

Extrauterine pelvic hemangiopericytomas presenting with LUTS and obstruction are extremely rare in the urologic setting. As in this case, while clinical diagnosis may not be immediately apparent, oncologic surgical principles regarding mass excision should be strictly followed for optimal outcomes. In conclusion, although rare, hemangiopericytomas should remain on a clinician's differential diagnosis when a patient presents with abdominopelvic pain due to an extrauterine pelvic mass and symptoms associated with compression of surrounding structures. Even if the mass appears radiographically benign and biopsy suggests an epithelioid mesenchymal neoplasm, it should be aggressively resected given the potential for malignant transformation, and the

patient must be followed closely to evaluate for recurrence and metastasis.

### Declaration of competing interest

The authors have no relevant conflicts of interest to declare.

### Acknowledgements

None.

### ABBREVIATIONS

CT	Computed tomography
eGFR	Estimated glomerular filtration rate
FNCLCC	Fédération Nationale des Centres de Lutte Contre le Cancer
LUTS	Lower urinary tract symptoms
MRI	Magnetic resonance imaging
POD	Post-operative day

### Consent

The subject provided written informed consent to the authors for publication of this case report and associated images.

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None.

### Author contributions

APM: Conceptualization; Patient treatment; Methodology; Data curation; Writing– Original draft preparation. MM: Data curation; Writing– Original draft preparation. DEZ: Patient treatment; Writing– Reviewing and editing. AKS: Conceptualization; Patient treatment; Supervision; Writing– Reviewing and editing.

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