# Giant Pelvic Schwannoma in the Setting of Renal Cell Carcinoma

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## Introduction

- Schwannomas are benign peripheral nerve sheath tumors composed of Schwann cells that typically occur in the head, neck, and extremities.[1] They are thought to be caused by a mutation in the NF2 gene that leads to improper functioning of the merlin suppressor protein.[2]
- Pelvic schwannomas are very rare accounting for about 1-3% of all schwannomas.[3] Once greater than 4.5cm, these tumors are classified as giant schwannomas.[4]
- Peak incidence occurs between age 20-50 with males and females equally affected.[3]
- They are difficult to diagnose as patients are asymptomatic until the tumor is large enough to compress surrounding structures leading to non-specific symptoms.[4-7]
- Official diagnosis can only be made after histopathological analysis. Pre-operative biopsy is an option but poses the risk of seeding if the tumor is malignant.[1]
- Surgical resection of these tumors is the treatment of choice.[1,3-7]
- Hypothesized that mutations in NF2 gene and SMARCB1 gene link the formation of schwannoma in neurofibromatosis type II with papillary renal cell carcinoma (RCC) and collecting duct RCC.[2,8,9]

## Case Description

**History:**

- 46 y/o male presented to the general surgery team with back/pelvic pain, hematuria, and pelvic mass of unknown origin for surgical consult.
- Denied fever/chills, night sweats, unexpected weight change, numbness/tingling of lower extremities, flank pain, constipation, or urinary symptoms.
- PMH: RCC, hypertension, hypothyroidism, diabetes mellitus type II, gastroesophageal reflux disease, and benign prostatic hyperplasia, attention-deficit hyperactivity disorder.
- RCC being monitored by urology and oncology through active surveillance.
- Medications: atenolol, lisinopril, metformin, levothyroxine, omeprazole, tamsulosin, tadalafil, alprazolam, amphetamine/dextroamphetamine.
- Allergies: Morphine.
- Past surgical history: None.
- Unremarkable social and family history.

**Physical Exam:**

Vitals:

- 156/82 mmHg, 95 bpm, 20 breaths/minute, 98.8F, 97% on room air.

Pre-operative:

- All systems unremarkable.

Post-operative:

- No acute distress, resting comfortably in the room.
- Breath sounds clear and equal bilaterally in all auscultatory fields.
- Regular rate and rhythm, S1&S2 appreciated with no murmurs, DP and PT pulses 2+ bilaterally.
- Abdomen soft, non-distended, appropriately tender to palpation over midline incision site
- Incision clean, dry, and intact with minimal serosanguineous staining on the dressing.
- A&O x4, lower extremity strength, motor function, and sensation all intact and equal bilaterally.

**Pre-Operative Differential Dx:**

- Liposarcoma, fibrosarcoma, ganglioneuroma, Tarlov cyst, and pelvic schwannoma.

**Labs/Diagnostics:**

- Figure 3 shows pre-operative CT of abdomen/pelvis with IV contrast which revealed 11 x 8.6 x 9.5cm pelvic mass and 2.7 millimeter enhancing right renal lesion.
- CBC and CMP within normal limits aside from white count to 18,400 $mm^3$ POD #1, which resolved by POD #2.
- Immunochemical stains performed by pathology included S100, SOX-10, Melan-A, and HMB-45.
- Figures 2 and 4 show specimen in pathology with final findings of spindle cell neoplasm consistent with schwannoma.

**Procedures:**

- Cystoscopy with ureteral stent placement was performed first to provide better visualization of the ureters intra-operatively.
- Open exploratory laparotomy revealed a large, well-encapsulated pelvic mass tightly adhered to S1, S2, and sacrum.
- R2 resection of pelvic mass was then performed to preserve the sacral plexus and patient's neurological function.

## Outcome

- Post-operative course was uncomplicated aside from hematuria, abdominal pain, and leukocytosis on POD #1, all of which resolved by POD #2.
- Patient was discharged on POD #3 after pain control was achieved, Jackson-Pratt drain was removed, and patient was tolerating regular diet with adequate bowel function.
- Patient was notified once the official pathology report was released, and follow-up was scheduled to monitor for recurrence.

## Discussion

- Complete surgical resection of pelvic schwannoma is the most effective treatment option with good prognosis post-operatively and low risk of recurrence.[1,3-7]
- Controversy exists surrounding whether preservation of neurologic function versus complete resection of the tumor is more important in cases where the tumor is adhered to the sacral nerve plexus.[3,7]
- Performing an R2 resection without knowing the histopathology of this tumor put our patient at higher risk of a poor outcome had this been a malignant tumor. This is a management problem that all providers face in pelvic masses of unknown origin.[1,3-6]

## Conclusion

- While there is not a well-studied genetic link between pelvic schwannoma and RCC, the case reports available show a strong potential for relationship between the two tumors.
- Pelvic schwannomas are difficult to diagnose and should be considered in the differential diagnosis of a patient with RCC exhibiting non-specific symptoms including low back pain, hematuria, or other urinary symptoms.

**Figure 1. Genetics of RCC and Schwannoma[8]**

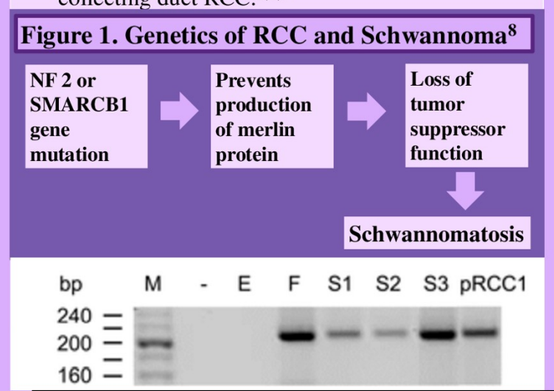


**Figure 2. Resected pelvic mass**

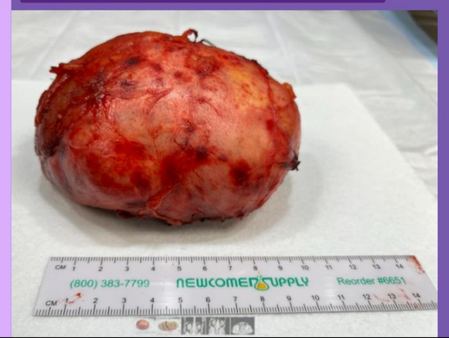


**Figure 3. Pre-op sagittal CT**

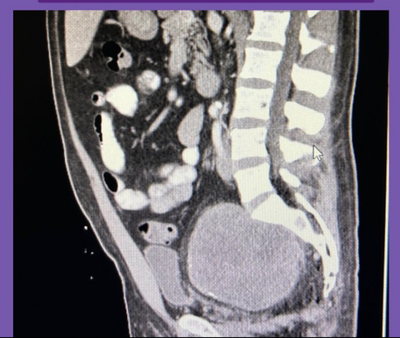

**Figure 4. Cross-sections of pelvic mass**


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