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Diagnosis and open excision of concurrent pelvic schwannoma and chromophobe renal cell carcinoma

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A B S T R A C T

Schwannomas are tumors that commonly affect the nerve sheath, typically involving peripheral and cranial nerves. These tumors are rarely found within the pelvis and retroperitoneum. To date, there have been no documented cases of concurrent chromophobe renal cell carcinoma and pelvic schwannoma. We present the case of a 57-year-old female with a right renal mass significant for chromophobe renal cell carcinoma and a left pelvic mass found to be a schwannoma. This case highlights the importance of adding schwannoma to the differential when considering a pelvic mass in the setting of renal cell carcinoma.

1. Introduction

Schwannomas are benign masses that form from Schwann cells of peripheral nerves and are classically associated with the 8th cranial nerve. The retroperitoneum and pelvis are a rare location for schwannomas with an estimated rate of 0.5 %–2 %.[1] While there have been several cases of pelvic, retroperitoneal, or renal schwannomas in the literature, there are no cases describing concurrent pelvic schwannoma and renal cell carcinoma (RCC) nor a description of management of schwannomas at the time of excision of malignant, solid tumors.[1,2] There has been a description of metachronous schwannoma and hybrid oncocytic/chromophobe tumors of the kidney, as well as papillary renal cell carcinoma suggesting there is a possible genetic relationship between renal masses and schwannomas. There may be a common pathogenesis to develop these tumors, and urologic surgeons should be aware of these relationships and options for management.[3,4] Therefore, we describe the diagnosis and multidisciplinary treatment of biopsy proven chromophobe RCC and pelvic schwannoma.

2. Case presentation

A 57-year-old female presented with a 9.6 × 10.1 cm right renal mass and a 6.7 × 8.2 cm left pelvic mass displacing her left ureter and iliac vessels. The patient underwent a percutaneous biopsy of both masses at an outside facility. Biopsy cores of the right renal mass were consistent with an eosinophilic chromophobe renal cell carcinoma (RCC) positive for PAX8, CK7, P504S and negative for MelanA. The left pelvic mass biopsy specimen was reportedly a benign fibroma on analysis of the initial biopsy specimen.

Initial imaging studies included a staging chest abdominal and pelvic CT angiogram with delayed venous phase which revealed a large right renal mass causing splaying of hilar vessels without evidence of renal vein invasion. Due to the significant vascular compression from mass effect of the pelvic mass, the patient was subsequently started on Apixaban prophylactically to prevent deep vein thrombosis. Re-interpretation by pathology of the left pelvic mass was consistent with a benign neurofibroma causing compression and near obliteration of left external and common iliac veins. Additionally, the mass resulted in severe narrowing of distal inferior vena cava and right common iliac vein. During history and physical exam, patient reported new onset symptoms of lower extremity peripheral artery disease likely secondary to this compression. Delayed venous imaging confirmed displacement of right common iliac and common iliac veins (image 1). No evidence of locally invasive or metastatic disease was identified on staging imaging. The patient's preoperative lab values are demonstrated in Table 1.

The patient was offered open right radical nephrectomy, cystoscopy with left ureteral stent placement and excision of the left pelvic mass with possible left iliac graft. Despite the challenges related to the tumor size and location, as well as extensive adhesions from previous surgeries, both the pelvic mass and kidney were excised successfully. The surgical procedure was completed in a multidisciplinary fashion, utilizing urologic, vascular, and neurosurgical teams. Due to the posterior adhesions

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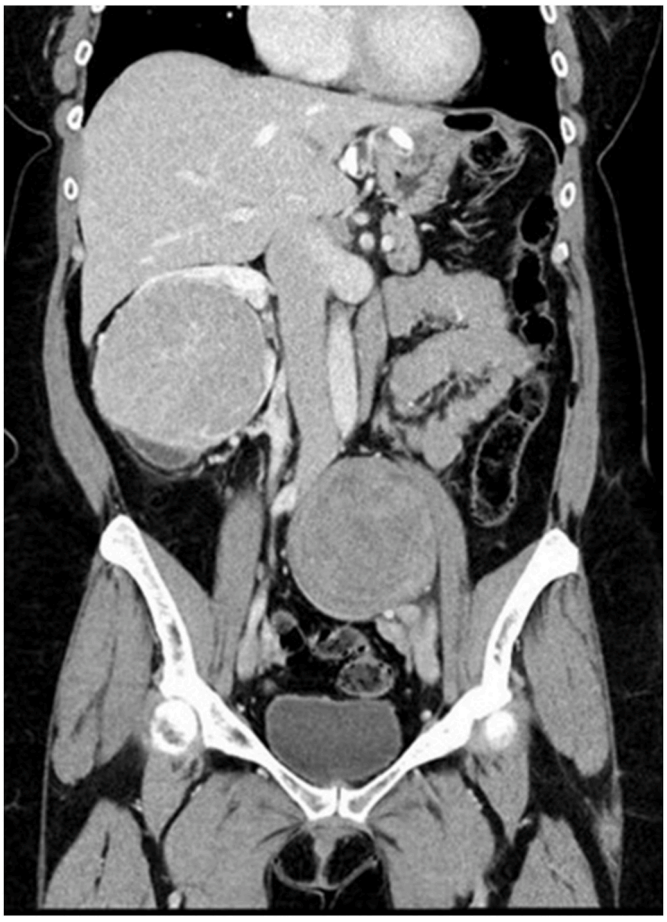

Image 1. Coronal view of CT Angiogram with delayed venous phase of abdomen and pelvis demonstrating splitting of left external iliac and common iliac veins.

Table 1
Patient's initial lab values.

Lab Value Marker	Patient's Value	Reference Value
Hemoglobin (Hb)	11.8 g/dL	13.0–16.5 g/dL
Hematocrit (Hct)	37.6 %	38.8–50.5 %
White Blood Cell Count (WBC)	4.3×10^3 cells/mm^3	$4.0–10 \times 10^3$ cells/mm^3
Platelet Count (PLT)	365×10^3 cells/mm^3	$150–450 \times 10^3$ cells/mm^3
Creatinine	1.02 mg/dL	0.5–1.2 mg/dL
Estimated GFR (eGFR)	64 mL/min/1.73m^2	>59 mL/min/1.73m^2

of the schwannoma to the lumbar spine and sacrum, neurosurgery assisted in the schwannoma excision. Vascular surgery assistance was required for a left common iliac vein graft, which was adherent to the pelvic mass and unroofed upon removal of the left pelvic mass.

The gross and microscopic pathology analysis of the right renal mass confirmed the diagnosis of chromophobe RCC (image 2 and 3) and the left pelvic mass was identified as a schwannoma that stained positive for S-100. Both masses were removed with negative margins. The excised schwannoma with intact capsule and microscopic evaluation of the mass is shown of image 3 and 4, respectively. The pathological stage classification of the right chromophobe RCC was pT2b (pTNM, AJCC 8th edition).

3. Discussion

Schwannomas are one of the most common tumors involving peripheral nerve sheaths. This category of tumors encompasses

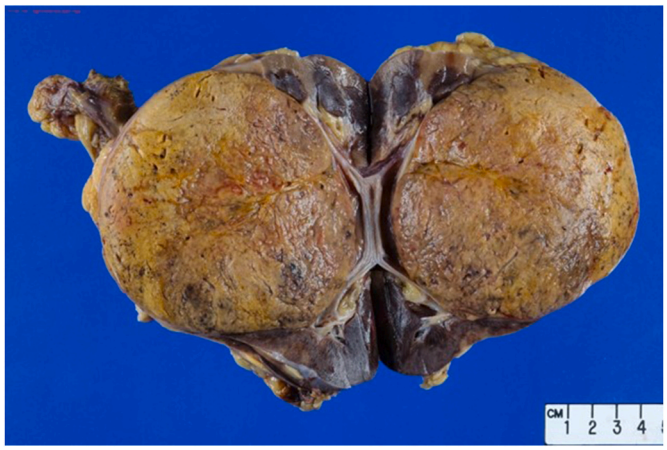

Image 2. Gross examination of excised right kidney; stage pT2b chromophobe renal cell carcinoma 12 cm in largest dimension with negative margins.

neurofibromas, perineuriomas, granular cell tumors and malignant peripheral nerve sheath tumors.[5] The cause of schwannomas is currently unknown.[5] There are very few documented cases of pelvic schwannoma and they typically arise from the sacral nerve or hypogastric plexus.[6,7] Pelvic schwannomas are predominately benign, but can be malignant in cases of von Recklinghausen's disease.[7]

Schwannomas are often asymptomatic until their growth leads to pain and/or compression of nearby structures usually leading to their discovery. When symptomatic, schwannomas can cause non-specific findings including back pain, abdominal or pelvic heaviness, symptoms of distension and discomfort.[6] Although imaging may assist in identifying a mass, definitive diagnosis usually relies on histological and immunohistochemical analysis of the specimen.[8] Upon immunohistologic analysis, cells are S-100 positive, confirming their neural crest origin, as seen in our patient.[9] In our case, immunohistochemical testing was key in distinguishing the schwannoma from the initially suspected fibroma. This testing was not performed on initial biopsy. Histologic examination of schwannomas reveals spindle cells with eosinophilic cytoplasm and elongated nuclei. These masses are typically encapsulated and organized in an Antoni A or Antoni B formation.[10]

The presence of chromophobe RCC in our patient with a large, concurrent pelvic schwannoma presented a significant surgical challenge. The presence of both masses required extensive diagnostic and presurgical planning. Understanding when to employ a multidisciplinary approach was critical. In our case, the pelvic schwannoma's size, orientation, and vascular involvement required vascular and neurosurgical assistance. It is important to be cognizant of these factors when considering surgical treatment. Current literature lacks specific information on outcomes related to patients with pelvic schwannoma and coexisting solid renal malignancy. There is limited evidence that suggests a genetic connection between renal masses and schwannomas, and we add to a growing amount of literature suggesting the presentations of these tumors may be more than simply sporadic. [3.4] This underscores the importance for further examination and documentation of these cases. While rare, pelvic schwannomas should be considered in patients with concurrent renal masses and complete excision at time of surgery can be considered.

4. Conclusion

We report a case of an incidentally discovered retroperitoneal schwannoma during staging of primary chromophobe RCC with synchronous surgical excision. Recent evidence suggests a genetic connection or predisposition of developing renal masses in patients with schwannomas. Although schwannomas are benign, larger extracranial lesions occasionally require excision due to compromise of adjacent

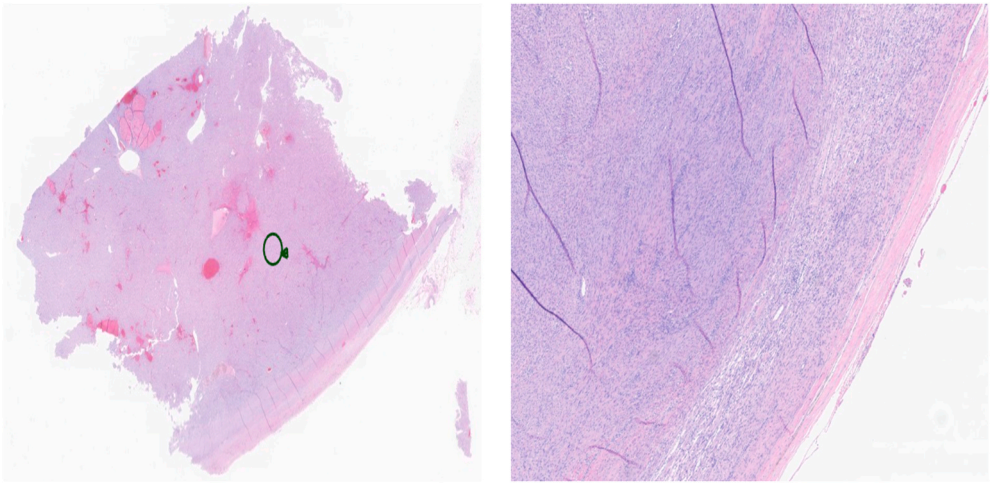

Image 3. Left: 0.5× magnification chromophobe renal cell carcinoma with overlying renal capsule and nested architecture. Right. 2× magnification schwannoma with capsule and underlying mass with diffuse spindle cells.

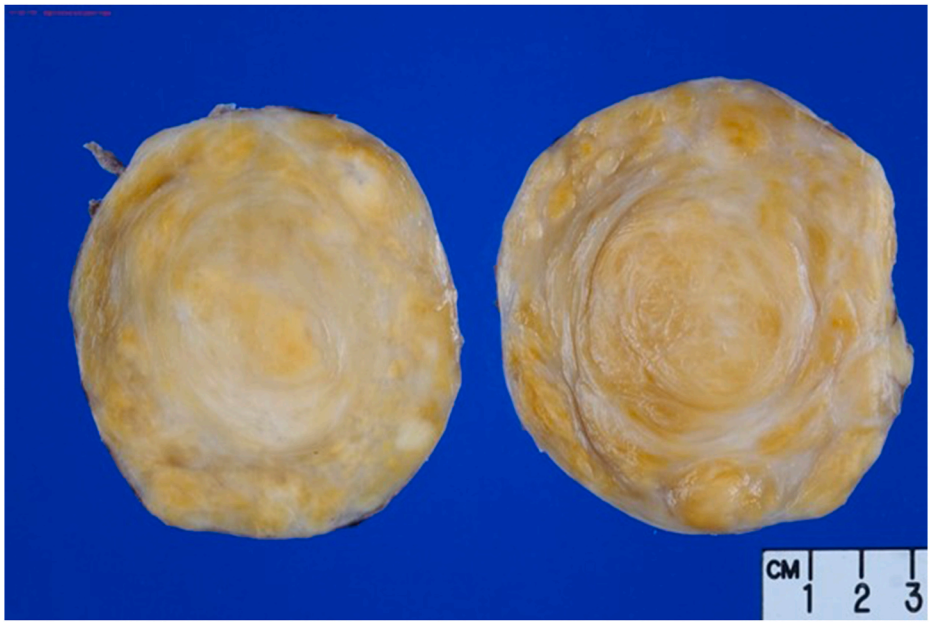

Image 4. Gross examination of excised left pelvic mass and schwannoma with negative margins.

organs or vascular structures. This required a multidisciplinary approach, first of its kind reported, and in this case, was accomplished with an excellent outcome.

CRediT authorship contribution statement

Charles Klose: Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Software, Resources, Project administration, Methodology, Investigation, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Mackenzie Gibbs:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Software, Resources, Project administration, Methodology, Investigation, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Amanda Kahn:** Writing – review & editing, Writing – original draft. **Bryce Baird:** Writing – review & editing, Writing – original draft, Methodology. **Sam Farres:** Validation, Supervision, Resources, Project administration, Methodology, Conceptualization. **Andrew Zganjar:** Writing – review & editing, Supervision, Project administration.

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