Background

- Synovial sarcoma (SS) is a malignant mesenchymal tumor that accounts for 14% of adult soft tissue sarcomas.
- SS mostly arises in juxta-articular locations within the deep soft tissue of the lower and upper extremities, particularly around the knee, which is the single most common location.
- Primary renal synovial sarcomas are exceedingly rare, representing approximately 1% of all renal malignancies.
- Due to its rarity and bland morphology, the diagnosis of synovial sarcoma in the urinary genital tract is often a challenge.
- The morphology, immunoprofiling and molecular studies, and diagnostic challenges of this case are discussed and the related literature is reviewed.

Methods/Materials

- The patient’s electronic medical records were thoroughly reviewed.
- Evaluation of CT imaging, renal biopsy, urine cytology and blood and pertinent laboratory testing.
- Literature was searched and reviewed using appropriate key words.

Case Report

- A 52-year-old male patient presented to an outpatient urology clinic with the main complaint of intermittent repeated gross hematuria and episodic left flank pain; urine analysis was positive for blood while his PSA and creatinine were within normal limits.
- CT imaging revealed a 2.5 cm enhancing solid mass near the left renal pelvis with hydrenephrosis.
- Given his clinical presentation and radiological findings, urothelial carcinoma was a concern.
- Initial non-invasive cytologic evaluation of voided urine and renal brush cytology revealed a predominantly hemorrhagic sample with a few clusters of atypical cells, favoring a reactive urothelial process.
- The final diagnosis of synovial sarcoma was only made on laparoscopic left nephroureterectomy.

Results

- The reported initial clinical presenting symptoms are non-specific including flank pain (approximately 32%), hematuria (30%), and less commonly lumbar pain, weight loss, and fever.
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- The clinical picture and radiologic findings of primary renal SS are entirely non-specific.

CT Abdomen and Pelvic: (A) Coronal CT imaging with and without contrast exhibited a marginally enhancing polyoid-like mass projecting into the lumen of the renal pelvis, measuring 2.5 cm in greatest dimension. (B) Maximum intensity projection studies showed limited contrast within the left upper pole collecting system and a 2.5 cm filling defect was noted within the renal pelvis of mixed density seen along the anterior aspect.

Histopathological Evaluation

- Hematoxylin and eosin-stained section of the tumor depicting a hypercellular, relatively well-circumscribed tumor, displaying a solid growth pattern and vague fascicular formations. The tumor is remarkably monotonous with minimal pleomorphism, originated in proximity to the renal pelvis. The cell margins are usually inconspicuous showing morphologic similarity with adjacent urothelial cells (top right), 20x.

- Tumor cells are monomorphic, with tight intermixing fascicles resembling a teratoma-like appearance. Minimal pleomorphism and mitotic activity were observed, 200x.

- Tumor cells are immunoreactive for PAX-8 and GATA-3, respectively, with variable nuclear intensity, 100x.

- Tumor cells are strongly and diffusely immunoreactive for TLE1, nuclear, 100x.

- SS18 (SYT) gene rearrangement detected by interphase FISH analysis, using break-apart FISH probes to detect rearrangements of the SS18 (SYT) gene at the 18q11.2 locus.

- Further research is required to determine whether GATA-3 is a predictive marker for synovial sarcomas.
- The clinical picture and radiologic findings of primary renal SS are entirely non-specific.
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Discussion/Conclusions

- Primary renal synovial sarcoma is a rare entity that shares a similar clinical and radiologic presentation with other renal malignancies, which is a challenge to the diagnosis and can be mislabeled.
- Upon literature review, primary renal synovial sarcoma ranged in size from 1.0 to 35.0 centimeters in greatest dimension with a mean of 10.4 centimeters (median 9.0 cm).
- The reported initial clinical presenting symptoms are non-specific including flank pain (approximately 32%), hematuria (30%), and less commonly lumbar pain, weight loss, and fever.
- GATA-3 positivity is reported to be associated with a worse prognosis in soft tissue sarcomas.
- Further research is required to determine whether TLE1 is a predictive marker for synovial sarcomas.

- Awareness of this entity, with the help of TLE1 and genetic tests, can diminish the challenge of this diagnosis.

References
