Renal sinus angiomyolipoma: A rare case

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INTRODUCTION

Angiomyolipoma (AML) consists of <10% of all renal tumors and is the most common benign mesenchymal neoplasm of the kidney.\(^1\) It is composed of varying amounts of poorly organized blood vessels, smooth muscles, and varying amount of fat.\(^2\) These tumors classically arise from the cortex and extend outward into perirenal fat. It is extremely rare occurrence for this tumor to arise from renal sinus and cause compression on the renal pelvis. Classical AML is easily diagnosed on cross-sectional imaging; however, fat-poor variant cannot yet be definitively diagnosed and differentiated from renal cell carcinoma (RCC).\(^2\) In our present case, renal sinus AML presented with hematuria and underwent radical nephroureterectomy as it could not be differentiated from transitional cell carcinoma (TCC) on imaging. After a thorough review, there are only 16 reported cases of renal sinus AML, this being the first case from India to our knowledge.

CASE REPORT

A 28-year-old female presented with dull aching pain in her right flank for one and a half months along with two episodes of gross hematuria with the passage of worm-like clots. There was no history of dysuria, fatigue, fever, or decreased urinary output. The patient did not suffer from any chronic medical ailment or bleeding disorders. There is no history of tobacco consumption in any form. Physical examination revealed no significant abnormality. Routine laboratory investigations were within normal limits. Urinalysis revealed red blood cell 100–200/HPF, pus cells 2–3/HPF, negative for nitrites, and leukocyte esterase. Urine cytology for malignant cells was negative. Ultrasonography suggested a focal hypoechoic lesion 25 mm × 20 mm in mid pole of the right kidney extending into renal pelvis. The color flow was seen in the lesion. Internal echos were present in pelvicalyceal system and upper ureter. Computed tomography (CT) urography reported an ill-defined hyperdense lesion, 23 mm × 18 mm × 23 mm, in right renal pelvis showing heterogeneous enhancement on postcontrast scans (Hounsfield unit [HU] ~80) with associated moderate dilatation of the right pelvicalyceal system suggestive of TCC or vascular malformation [Figures 1-3].

Keywords: Angiomyolipoma, rare, renal sinus

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With a suspicion of TCC, the right nephroureterectomy was performed. Biopsy report was grossly suggestive of well-circumscribed submucosal renal pelvic mass 2.5 cm × 2cm × 1.5 cm. On microscopy, sections of mass showed a tumor predominantly composed of mature adipose tissue, scattered blood vessels, and proliferated lymphoplasmacytic cells. It also showed spindle cells which on immunohistochemistry stained negative for desmin, H-Caldesmon and Anaplastic lymphoma kinase (ALK) but took red stain with Masson trichrome stain suggestive of smooth muscle cells.

**DISCUSSION**

AML is the most common benign mesenchymal neoplasm of the kidney. It is composed of varying amounts of poorly organized blood vessels, smooth muscles, and varying amount of fat. These constitute of <10% of all renal tumors. Although initially considered a form of hamartoma, they are now considered to belong to a family of neoplasms called perivascular epithelioid cell tumors (“PEComas”).

Most AML occurs sporadically with 20%–30% cases associated with tuberous sclerosis. Two AML most commonly present in middle-aged females as flank pain and hematuria. They are also the most common neoplasms associated with spontaneous hemorrhage, the risk of which increase as the size of tumor increases.

These tumors usually arise in the renal cortex and expand into the perirenal fat. It is extremely rare for this tumor to arise from the renal sinus and cause compression of the renal pelvis. Classical AML is easily diagnosed on cross-sectional imaging; however, fat-poor variant cannot yet be definitively diagnosed and differentiated from RCC. On noncontrast CT, AML appears as well-circumscribed mass showing areas of hypo attenuation (−20 HU) which is typical of these tumors. However, fat poor AML, occurring in 5% cases, differentiation from other solid masses is less likely. In our case, imaging characteristics were not typical of AML and along with its rare location, the possibility of TCC could not be ruled out.

Tumors of the renal sinus consist of lipomas, leiomyomas, AMLs, etc., and the manifestations are nonspecific. Patients may present with flank pain and gross hematuria specially when the size exceeds 4 cm. Symptomatic tumors mandate resection; however, renal preservation should be considered as far as possible considering the benign nature of the disease. In these centrally located tumors, near the renal hilum nephron-sparing surgeries are technically more demanding with increased incidence of longer ischemia times, collecting system violation, and overall complications. In our case, the pain might have been caused due to compression of the pelvicalyceal system causing its dilatation. In the present study, the patient underwent radical nephroureterectomy with suspicion of TCC of the renal pelvis.

To best of our knowledge, only 16 cases of AML originating from renal sinus have been previously reported and in most of the reports, these underwent a radical surgery due to uncertainty of the nature of the lesion.

In conclusion, AML presenting as renal pelvic masses are extremely rare with only a few cases been reported till date. They are most often misdiagnosed as renal pelvic TCC. The prognosis of sporadic AML is good. With more advanced CT and multiparametric magnetic resonance imaging protocols orientation and qualitative diagnosis of these tumors are expected to improve. Preserving the kidney
during surgery as far as possible is key for the treatment of patients with such tumors. However, rarity of the tumor, along with difficulty in diagnosing based on the current imaging studies, makes nephron-sparing surgery relatively under-utilized.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Eble JN, Sauter G, Epstein JI, Sesterhenn IA. World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of the Urinary System and Male Genital Organs. Lyon, France: International Agency for Research on Cancer; 2004.
2. Margulis V, Karam JA, Matin SF, Wood CG. Benign renal tumors. In: Wein AJ, editor. Campbell–Walsh Urology. 11th ed. Philadelphia: Elsevier; 2016. p. 1306-9.
3. Shirotake S, Yoshimura I, Kosaka T, Matsuzaki S. A case of angiomyolipoma of the renal sinus. Clin Exp Nephrol 2011;15:953-6.
4. Martorana G, Lupo S, Brunocilla E, Concetti S, Malizia M, Vece E. Role of nephron sparing surgery in the treatment of centrally located renal tumors. Arch Ital Urol Androl 2004;76:51-5.
5. Cheng B, Cai Q, Wu Y, Zhao Y, Guo Q, Li G, et al. Primary renal sinus tumor: Three case reports with a review of the literature. Oncol Lett 2015;9:829-32.