Angiomyolipoma Lesion Causing Unilateral Ureteral Obstruction

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Citation

Abstract
A 22-year old female with known tuberous sclerosis presented with right flank pain and was found to have right ureteral obstruction due to a lower pole angiomyolipoma. Selective angio-embolization of the lesion was unsuccessful and the patient underwent right partial nephrectomy. The patient had an uneventful post-operative course and experienced resolution of her flank pain. Patients with known angiomyolipomas must be followed closely to monitor for renal obstruction so that early intervention may be pursued when necessary.

INTRODUCTION
Angiomyolipomas identified in patients with the tuberous sclerosis are typically multicentric, bilateral, and tend to enlarge significantly over time. Although histologically benign, these lesions can rupture spontaneously and result in retroperitoneal hemorrhage. Ureteral obstruction associated with angiomyolipoma is rare. We report a case of unilateral ureteral obstruction and hydronephrosis caused by a lower pole angiomyolipoma. To our knowledge, hydronephrosis due to angiomyolipoma has not been previously reported.

CASE REPORT
A 22-year old female with a known history of tuberous sclerosis and bilateral angiomyolipomas presented with right flank pain. Laboratory evaluation revealed a serum creatinine of 0.6mg/dL. An infused CT scan of the abdomen and pelvis identified numerous, bilateral hypodense lesions. Imaging revealed an adipose component to these lesions that was consistent with angiomyolipoma. The largest lesion was a 5.8 x 3.8 cm, exophytic mass located on the inferomedial aspect of the right kidney. This lesion compressed the ureter and resulted in moderate hydronephrosis (Figure 1). The patient underwent selective angio-embolization of the lesion. A third-order renal artery branch was identified as the main arterial supply to the obstructing angiomyolipoma and coil embolization was performed. Continued hydronephrosis was noted on follow-up imaging and the patient underwent ureteral stent placement for temporary relief of the obstruction.

One month following attempted embolization, the patient underwent a right partial nephrectomy. The kidney was observed to be large and associated with an inflammatory and desmoplastic reaction thought to result from the prior procedures. Following careful dissection, the ureter was identified and observed to be completely encased within the lower pole angiomyolipoma (Figure 2a). Then tumor was then dissected free from the ureter (Figure2b) and sent to
pathology for tissue confirmation. The ureteral stent was left in place situ and removed two weeks postoperatively during routine follow-up.

Figure 2
Figure 2: Intra-operative photo of (A) lower pole tumor encasing the ureter and (B) the ureter following dissection free from the tumor.

**DISCUSSION**

Angiomyolipomas are benign lesions that are usually asymptomatic. The majority of these lesions do not require surgical intervention. Flank pain, due to localized hemorrhage and/or inflammation, is both the most common presenting symptom and indication for intervention.2 Historically, management of angiomyolipomas has been based on tumor size and the presence or absence of clinical symptoms. In patients with tuberous sclerosis, preservation of renal function is imperative. For this reason, conservative management of asymptomatic tumors is often advocated. Nephron-sparing procedures such as selective angi-embolization and open partial nephrectomy have been successfully reported in the literature.3 Although patients with angiomyolipomas may develop renal insufficiency or failure due to chronic compression and replacement of normal renal parenchyma by angiomyolipoma lesions, ureteral obstruction has not been described. We report the use of partial nephrectomy in the treatment ureteral obstruction resulting from angiomyolipoma. In patients with known angiomyolipomas, we recommend routine imaging in order to detect early signs of obstruction prior to the onset of irreversible renal damage.

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