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Reed Ritterbusch

Providence, reed.ritterbusch@providence.org

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Spontaneous Rupture of a Previously Asymptomatic Giant Angiomyolipoma

Reed Ritterbusch, MD

Introduction

- Renal angiomyolipoma (AML) is a benign mesenchymal neoplasm in the kidney with an incidence of around 0.13% in the population, with a higher prevalence in women. About 80% occur sporadically while 20% are associated with tuberous sclerosis.
- Most are asymptomatic with incidental diagnosis; however, large AMLs of >4cm can present symptomatically with pain, palpable mass, mass-associated symptoms, or hemorrhage.
- Giant AMLs, defined as those with diameter >10cm are especially uncommon with only a handful described in the literature, and are prone to hemorrhage.

Learning Objectives

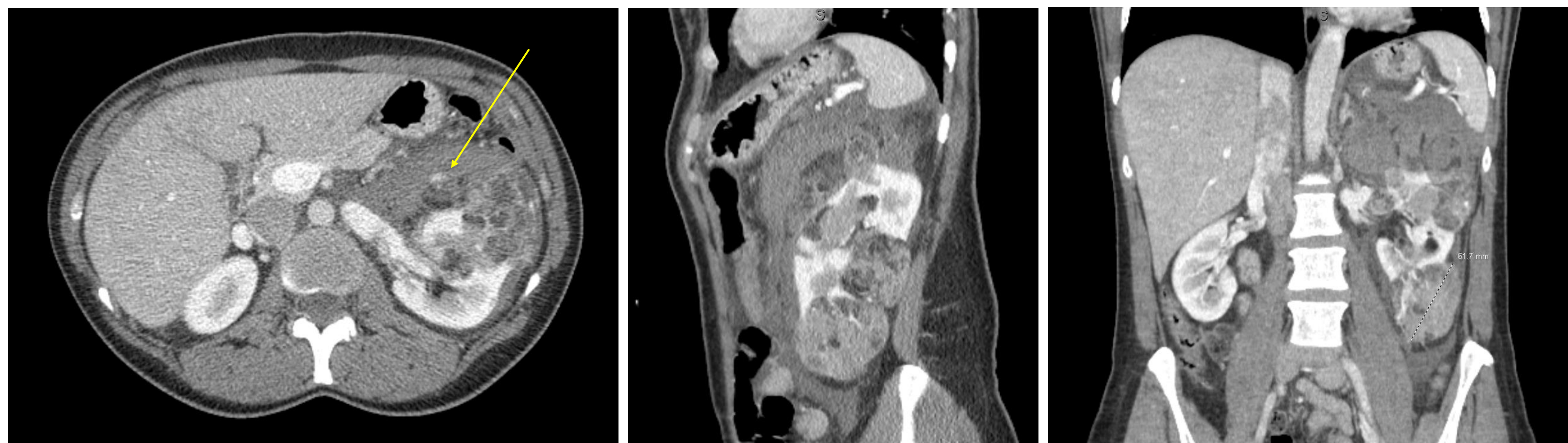
- Rare case of multiple ruptured angiomyolipomas, including a giant angiomyolipoma, exhibiting both Lenk's triad as well as Wunderlich's syndrome
- Lenk's Triad: acute flank pain, abdominal tenderness and signs of internal bleeding
- Wunderlich's syndrome: Spontaneous renal hemorrhage
- Case differential initially included gastroenteritis, diverticulitis, ovarian torsion, kidney stone
- Prompt CT imaging showed the correct diagnosis, and she was able to be stabilized with renal artery embolization with later definitive treatment of left nephrectomy.

Case Presentation

- History:** Patient is a 49-year-old female who presented to the emergency department with abdominal pain.
- 9/10, squeezing, constant, worse with movement, in the LLQ
- Non-bloody diarrhea, chills, and nausea. Denied fever, vomiting, hematuria
- Eaten sushi two days prior. No recent antibiotic use.
- PMH: History of kidney stones. PSH: appendectomy
- Exam:** notable for blood pressure of 96/52 and tenderness to palpation in the left lower quadrant and left costovertebral angle.
- Labs:** WBC 16.0. H/H 12.0/35.8. Platelets 246. BUN 12. Creatinine 0.72. Lipase within normal limits. Negative pregnancy test. Liver function tests within normal limits.

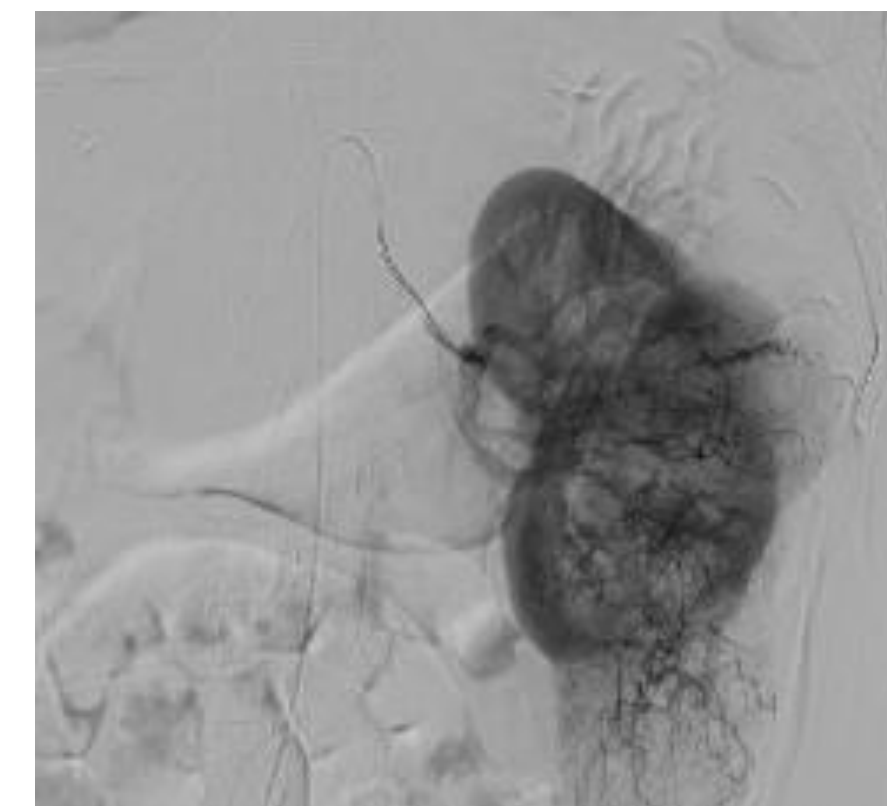
Diagnostic Imaging

CT abdomen and pelvis with contrast showed multiple left renal AMLs with acute hemorrhage involving a mass at the upper pole of the kidney with active extravasation of contrast. A large retroperitoneal hematoma exhibited mass effect on surrounding structures.



Intervention

- Patient was taken to interventional radiology where a left renal angiogram and bead embolization of two AMLs were performed.
- Following the procedure, she remained hemodynamically stable and was discharged with follow up with urology.
- She later underwent an elective robotic assisted nephrectomy
- Pathology showed multiple AMLs, with the largest being 14.5 cm in greatest dimension.



Discussion

- This is a rare case of spontaneous rupture of previously asymptomatic angiomyolipomas, including a giant (>10cm) angiomyolipoma with retroperitoneal hemorrhage with associated mass effect
- In contrast to other benign renal masses, the diagnosis can be made on imaging by the presence of intralesional fat (-15 to -20 Hounsfield units)
- Wunderlich's syndrome: spontaneous renal hemorrhage, which is a highly uncommon but life-threatening urological emergency that can occur with AMLs greater than 4 cm
- About 20% of those presenting with symptomatic AMLs will have renal hemorrhage
- Lenk's triad: acute flank pain, abdominal tenderness, and signs of internal bleeding such as hypotension or hematuria
- Management options include observation for tumors less than 4cm and embolization or partial or total nephrectomy for those larger

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