Spontaneous Rupture of a Previously Asymptomatic Giant Angiomyolipoma

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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 (>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:** History of kidney stones.
- **Exam:** notable for blood pressure 9/10, squeezing, constant, worse with movement. Temperature 102.2°F, respiratory rate 20, heart rate 100

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 intrarenal 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

References