

# AMSER Case of the Month

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36-year-old female with right lower quadrant  
abdominal pain

Gabriel Hanson, M2

Matthew Kruse, M.D.

Creighton University School of Medicine

# Patient Presentation

**HPI:** A 36-year-old female presents to the ED with a chief complaint of severe right sided abdominal pain that is sharp in nature, constant, and radiating to the groin and back. Patient reports associated nausea.

**PMH:** Tuberous sclerosis

**PSH:** Cholecystectomy, Hysterectomy

**Physical Exam:** Right lower quadrant is tender to palpation

# Pertinent Labs

- **CBC:** WBC 17.3, HBG 11.8, ANC 12.8
- **UA:** Appearance – slightly cloudy, Blood – moderate, Protein – 30, RBC – 10-20, Casts Granular – 0-2, Amorphous – 2+, Squam Epithelial Cells – 5-10
- **CMP:** Glucose – 205, Creatinine – 1.23, Chloride – 113, CO2 – 19.0, GFR – 56
- **Lactic Acid, Venous** – 3.2

What Imaging Should We Order?

# ACR Appropriateness Criteria

## Variant 1:

Right lower quadrant pain. Initial imaging.

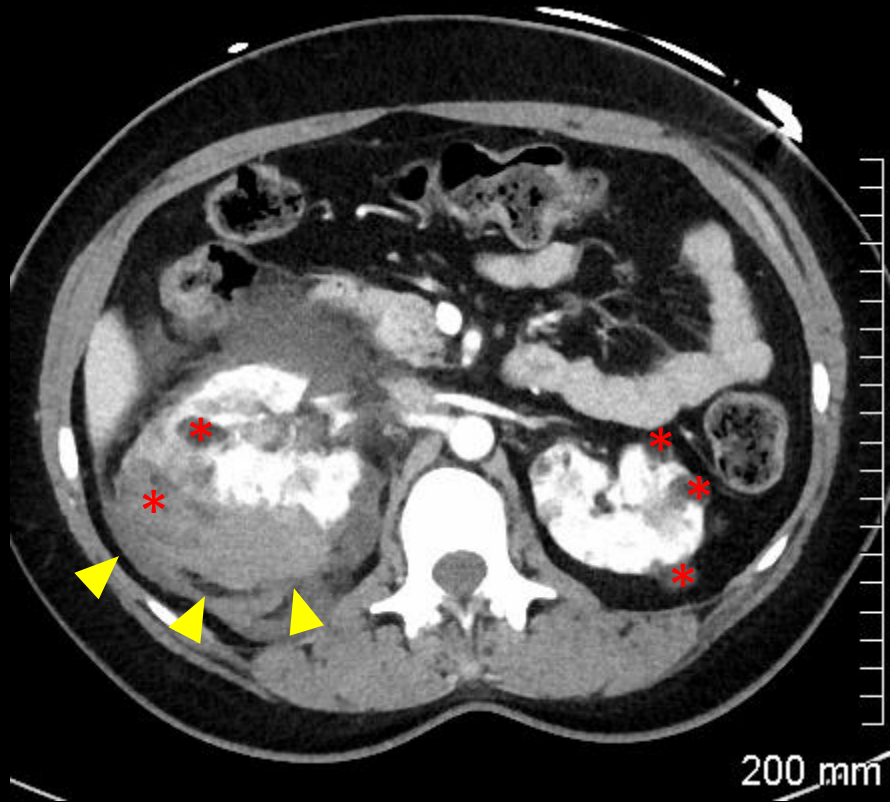
Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	⊕⊕⊕
US abdomen	May Be Appropriate	○
US pelvis	May Be Appropriate	○
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	○
MRI abdomen and pelvis without IV contrast	May Be Appropriate	○
CT abdomen and pelvis without IV contrast	May Be Appropriate	⊕⊕⊕
Radiography abdomen	Usually Not Appropriate	⊕⊕
Fluoroscopy contrast enema	Usually Not Appropriate	⊕⊕⊕
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
WBC scan abdomen and pelvis	Usually Not Appropriate	⊕⊕⊕⊕

This imaging modality was ordered by the ER physician

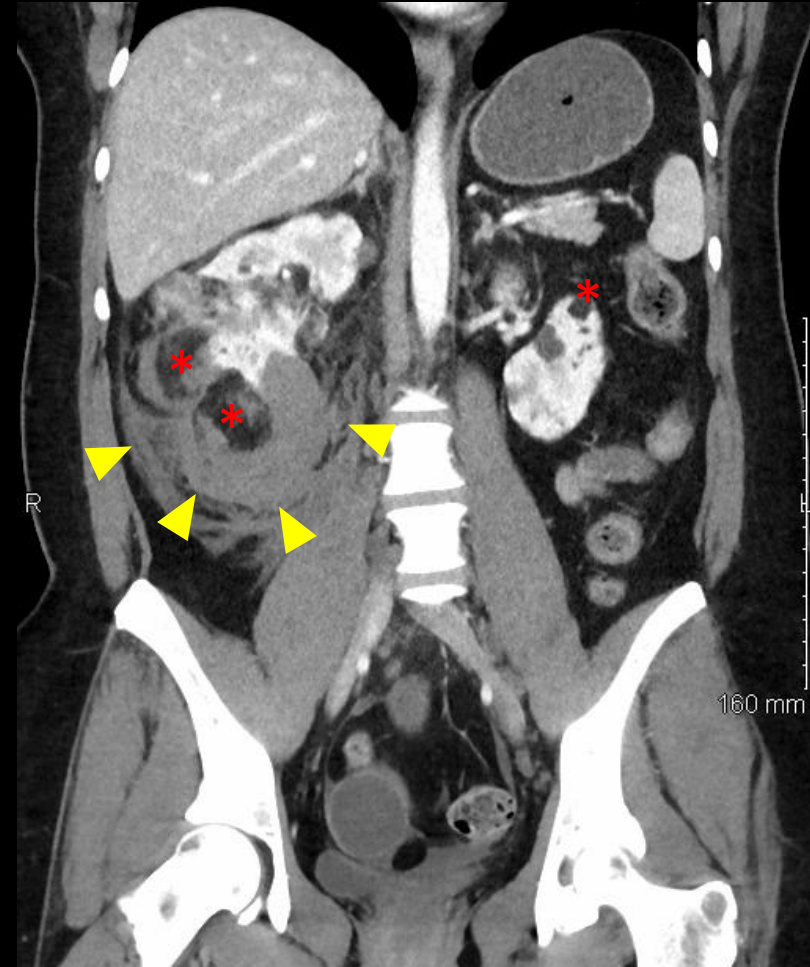
# CT with Contrast Findings



# CT with Contrast Findings (Labeled)



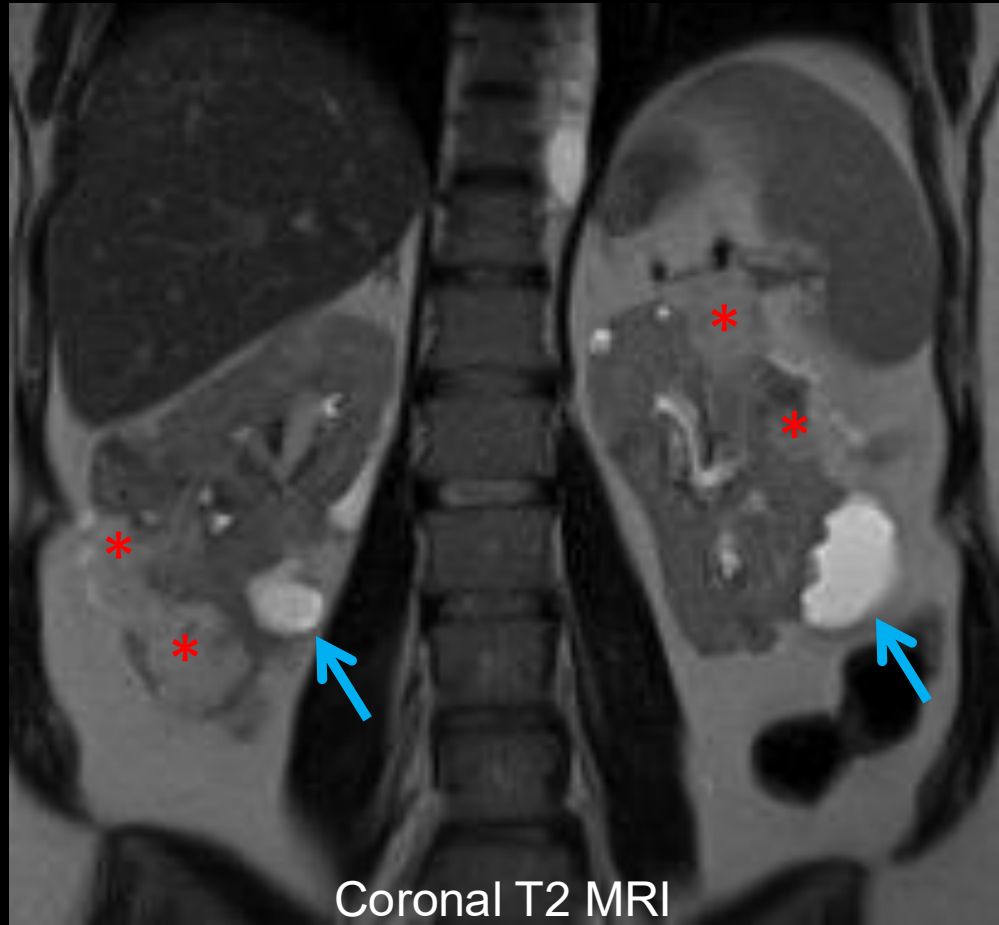
- Acute perirenal hemorrhage (>) from ruptured right angiomyolipoma



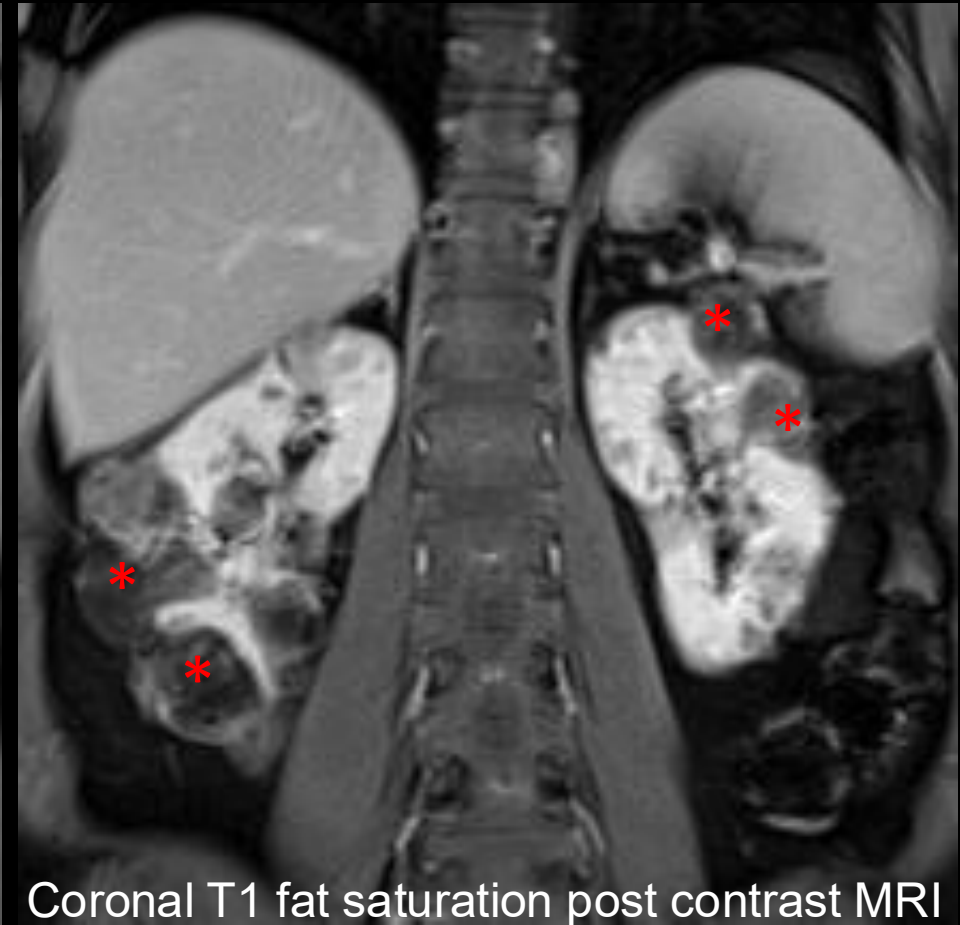
- Multiple bilateral fat-containing renal masses compatible with angiomyolipomas (\*)



# Previous MRI Abdomen with and without contrast



Multiple bilateral fat-containing renal masses compatible with angiomyolipomas (\*)



Renal cysts (→)



## Final Dx:

Ruptured right angiomyolipoma with acute  
perirenal hemorrhage in a patient with  
tuberous sclerosis

# Case Discussion

## Tuberous Sclerosis (TSC):

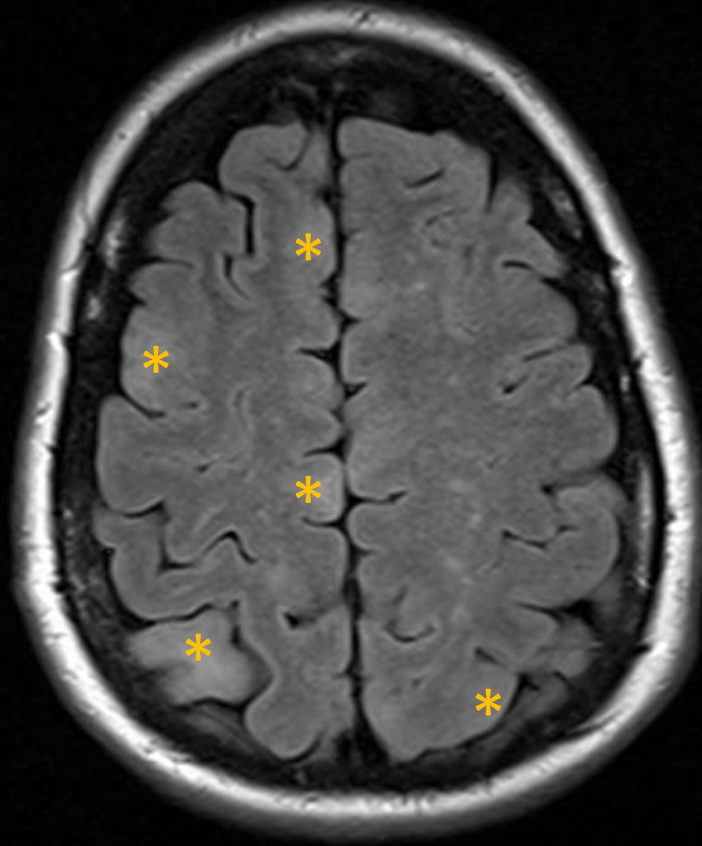
- Rare AD neurocutaneous disorder due to mutations in the TSC1 or TSC2 tumor suppressor genes [1].
- Classically diagnosed in children and presents with seizures, developmental delay, and hypomelanotic macules [2].
- Prevalence is estimated to be 1 in 6000 people [2].
- Renal failure is the leading cause of death in patients with TSC [3].

# Case Discussion

## TSC Imaging Manifestations [2]:

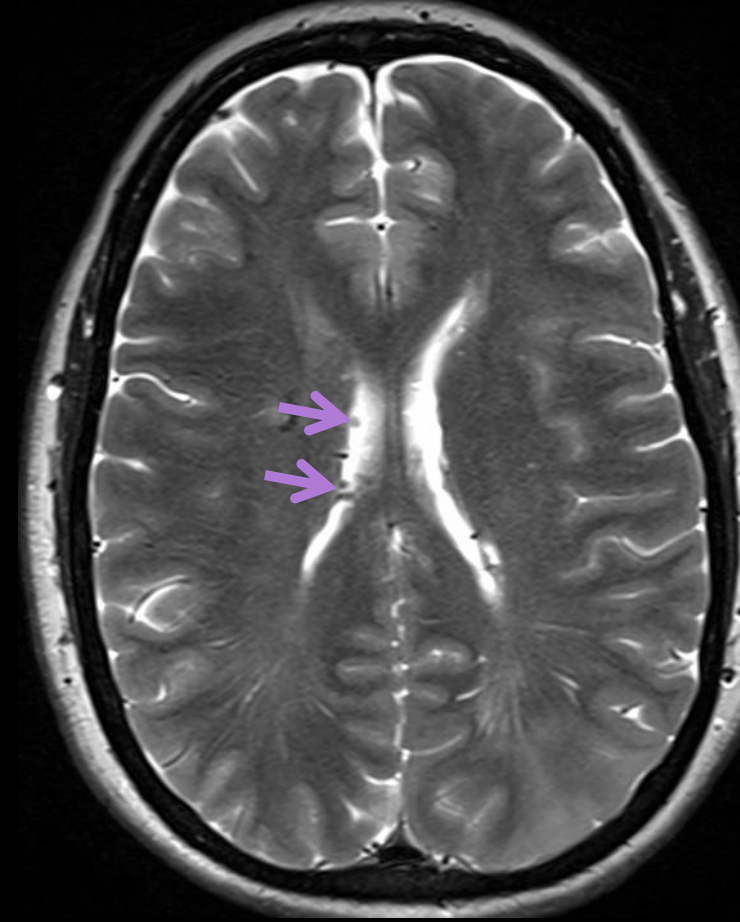
- Renal → angiomyolipomas, cysts, and renal cell carcinoma
- CNS → subependymal nodules, cortical/subcortical tubers, cerebral white matter radial migration lines, and subependymal giant cell astrocytomas
- Pulmonary → lymphangioleiomyomatosis
- Cardiac → rhabdomyomas
- Hepatic → angiomyolipomas
- Splenic → hamartomas

# Previous MRI Brain for our patient



Axial T2 FLAIR MRI

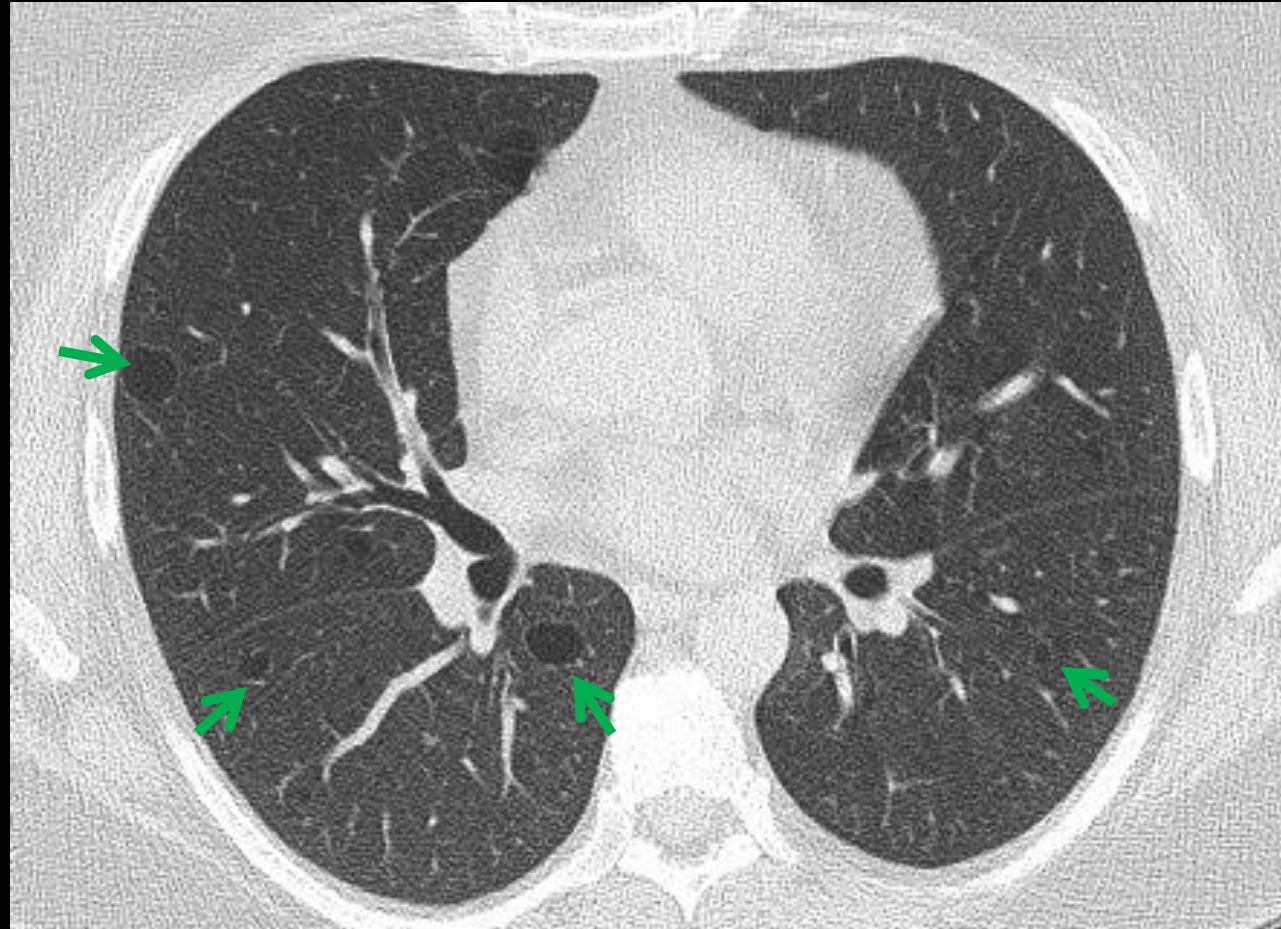
Multiple T2/FLAIR hyperintense cortical and subcortical tubers / hamartomas (\*)



Axial T2 MRI

Multiple small subependymal nodules (→) compatible with hamartomas

# Subsequent Noncontrast CT Chest for our patient



Multiple thin-walled round pulmonary cysts (→)  
compatible with lymphangioleiomyomatosis

# Case Discussion

## Renal Manifestations of TSC:

- Angiomyolipomas (AMLs)
  - Hamartomatous lesions consisting of abnormal blood vessels, smooth muscle, and adipose tissue [3].
  - Diagnosed as a renal mass containing macroscopic fat [2].
    - Although ~5% may not contain obvious fat
  - Contain micro/macroaneurysms that may spontaneously rupture and bleed [3].
  - Lesions >4 cm are at greater risk of spontaneous hemorrhage [3].
- Cysts
  - Commonly multiple and typically asymptomatic [2].
- Renal Cell Carcinoma
  - Incidence rate is similar to general population (2-3%), but age of onset is significantly lower [2].

# Case Discussion

Management of renal angiomyolipomas:

- Trending towards nephron-sparing strategies, referred to as active surveillance (AS) [4].
  - Allows for maximum preservation of renal structure and avoids risks associated with procedures [4].
  - mTOR inhibitors are becoming more frequently used as targeted therapy for AMLs that are >4cm [5].
- Active treatment (AT) options include surgical excision, angioembolization and cryoablation [4].
  - Specific situations in which AT is preferred include patients with large tumors (>4 cm), women of childbearing age, or patients who may have difficulties following up in the future [4].
  - Risk-benefit analysis should be performed prior to surgery [5].



# Case Discussion

- The patient's HGB remained stable after an initial drop, permitting continued active surveillance as the chosen conservative approach.
- Care team considered starting mTOR inhibitors to help reduce risk of bleeding and cytopenias.
- After a 3 day hospital stay and adequate pain management, the patient was discharged home with instructions to follow up with PCP in 1 week and Heme/Onc in ~4 weeks.

# References:

1. Wang MX, Segaran N, Bhalla S, et al. Tuberous Sclerosis: Current Update. *RadioGraphics*. 2021;41(7):1992-2010. doi:<https://doi.org/10.1148/rg.2021210103>
2. Manoukian SB, Kowal DJ. Comprehensive Imaging Manifestations of Tuberous Sclerosis. *American Journal of Roentgenology*. 2015;204(5):933-943. doi:<https://doi.org/10.2214/ajr.13.12235>
3. Casper KA, Donnelly LF, Chen B, Bissler JJ. Tuberous Sclerosis Complex: Renal Imaging Findings. *Radiology*. 2002;225(2):451-456. doi:<https://doi.org/10.1148/radiol.2252011584>
4. Daché A, Fatica R, Herts BR, et al. Factors predicting the active treatment of renal angiomyolipoma: 30 years of experience in two tertiary referral centers. *Frontiers in surgery*. 2023;10. doi:<https://doi.org/10.3389/fsurg.2023.1094806>
5. Wang H, Long Q, Wang Y, Liu L, Zhou L, Guo J. Tuberous sclerosis complex-associated renal angiomyolipomas: A single center study of 17 consecutive cases. *Oncology Letters*. 2016;12(2):1501-1506. doi:<https://doi.org/10.3892/ol.2016.4766>