Renal Angiomyolipoma with Spontaneous Retroperitoneal Hemorrhage in a Patient with Tuberous Sclerosis Complex

Tuberous Sclerosis Complex (TSC) is an uncommon genetic, multisystemic disorder with an incidence of approximately 1 in 5,000 to 10,000 live births, that results in the development of noncancerous tumors in different parts of the body (1). Common manifestations of TSC include renal angiomyolipomas, neurofibromas, pulmonary lymphangioleiomyomatosis (LAM), amongst others. Our patient was a 51-year-old female with a history of TSC and pulmonary LAM who presented with symptoms of dizziness and left-sided abdominal pain, without a history of trauma. Upon computed tomography (CT) imaging, she was found to have a large exophytic, spontaneously ruptured, renal angiomyolipoma arising from the lower pole of the left kidney with abundant retroperitoneal hemorrhage (Figure 1), consistent with Wunderlich Syndrome. CT also revealed multiple thin-walled pulmonary cysts distributed throughout both lungs and a small left pleural effusion, most compatible with pulmonary LAM (Figure 2). Once hemodynamic stabilization was accomplished, the patient underwent successful embolization.

Renal angiomyolipomas, previously classified as hamartomas, are the most prevalent benign renal tumors. They are generally highly vascular and primarily composed of smooth muscle and adipose tissue (2). Renal angiomyolipomas are present in 25% to 50% of TSC patients and are usually bilateral and multicentric. Most angiomyolipomas are found incidentally upon radiological imaging, however, symptomatic presentations also exist. Typical symptoms include flank pain and gross hematuria (2). A tumor with a diameter larger than 4 cm is more likely to develop aneurysms and is more susceptible to bleeding (3). Selective embolization of a renal angiomyolipoma is considered to be a safe and minimally invasive procedure with low complication rates. It is important to keep in mind the possibility of a renal angiomyolipoma in a patient with TSC and flank pain, and that if present, there can be spontaneous rupture even if there is no history of trauma. The availability of CT is essential for the evaluation and diagnosis of this condition. Early and accurate diagnosis is vital for effective management.

Víctor L. Guzmán-Rivera, MD, Diagnostic Radiology Resident PGY- 5;
Claudia M. Muns, MD, Diagnostic Radiology Resident PGY- 3;
Ivonne Ojeda, MD, Diagnostic Radiology Attending Physician;
Diagnostic Radiology, Department of Radiological Sciences, School of Medicine, University of Puerto Rico Medical Sciences Campus, San Juan, Puerto Rico.

References

1. Northrup H, Koenig MK, Pearson DA, Au KS. Tuberous Sclerosis Complex. 1999 Jul 13 [updated 2021 Dec 9]. In: Adam MP, Feldman J, Mirzaa GM, Pagon RA, Wallace SE, Bean LJH, Gripp KW, Amemiya A, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2023. PMID: 20301399.
2. Shamam YM, Leslie SW. Renal Angiomyolipoma. 2023 Aug 8. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan–. PMID: 36256751.
3. Azzi S, Jissendi P, Tannouri F. Ruptured Renal Angiomyolipoma. J Belg Soc Radiol. 2022 Nov 4;106(1):100. doi: 10.5334/jbsr.2932. PMID: 36382018; PMCID: PMC9635320.

Figures 1 and 2: 1) CT axial image of a left-sided renal angiomyolipoma, measuring 8.5 cm anteroposterior (AP) by 8.4 cm transverse by 9.6 cm craniocaudal (CC), with an associated retroperitoneal hemorrhage, consistent with Wunderlich Syndrome. 2) CT axial image of multiple thin-walled pulmonary cysts distributed throughout both lungs and a small left pleural effusion, most compatible with pulmonary LAM.