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Spontaneous Hemorrhage of a Renal Angiomyolipoma: Case Report and Review

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A B S T R A C T

Background: Angiomyolipoma is a benign renal tumor composed of fat tissue, smooth muscle, and vascular tissue. Angiomyolipoma tumors are usually asymptomatic and diagnosed incidentally. The tumor hemorrhage is the most common complication of angiomyolipoma. Moreover, Wunderlich syndrome is a rare spontaneous phenomenon associated with sub-capsular and perirenal hemorrhage with no history of trauma.
Case Report: This case report presents a spontaneous hemorrhage of a renal angiomyolipoma.
Conclusion: Angiomyolipoma can be presented with sub-capsular and perirenal hemorrhage with no history of trauma that can be life-threatening.

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Introduction

Angiomyolipoma is a benign tumor consisting of different proportions of adipose, smooth muscle, and abnormally thick-walled blood vessels. In 5% of the tumors, the fat portion of the tumor can only be detected on a microscopic scale (1). The prevalence rate of renal angiomyolipoma is from 0.3% up to 3%. This tumor originates from the renal mesenchymal tissue. In general, the prevalence rate of angiomyolipoma is four times higher in females, compared to males, which is due to the effect of hormones that are effective in tumor growth (2). The angiomyolipoma can present as a sporadic tumor or associated with tuberous sclerosis complex (3). Most angiomyolipomas are asymptomatic and diagnosed accidentally during an ultrasound or computed tomography (CT) scans. Symptomatic cases can be spontaneous hemorrhages that may cause the patient to experience hypovolemic shock and even death. The bleeding that leads to hypovolemic shock is called Wunderlich syndrome that presents with flank or abdominal pain, a palpable tender mass, and gross hematuria (Lenk's triad) (4, 5).

The risks of bleeding in tumors less and more than 4 cm in diameter are 13% and 51%, respectively (6, 7). A renal angiomyolipoma that grows more than 10 cm in size is called a giant angiomyolipoma. Few studies have reported tumors larger than 20 cm in this regard (8). However, due to bleeding, aneurysms are caused by the increasing size of the angiomyolipoma, pressure symptoms, and risk of bleeding owing to rupture increase (9). Giant angiomyolipoma usually presents with clear clinical symptoms, and the preservation of kidney function is the main goal of treatment in these cases (10). In this case study, we report a case of renal angiomyolipoma with spontaneous hemorrhage.

Case Report

A 67-year-old female presented to the emergency department with sudden left flank pain, weakness, nausea, vomiting, and hematuria. The pain was vague and persistent, which increased

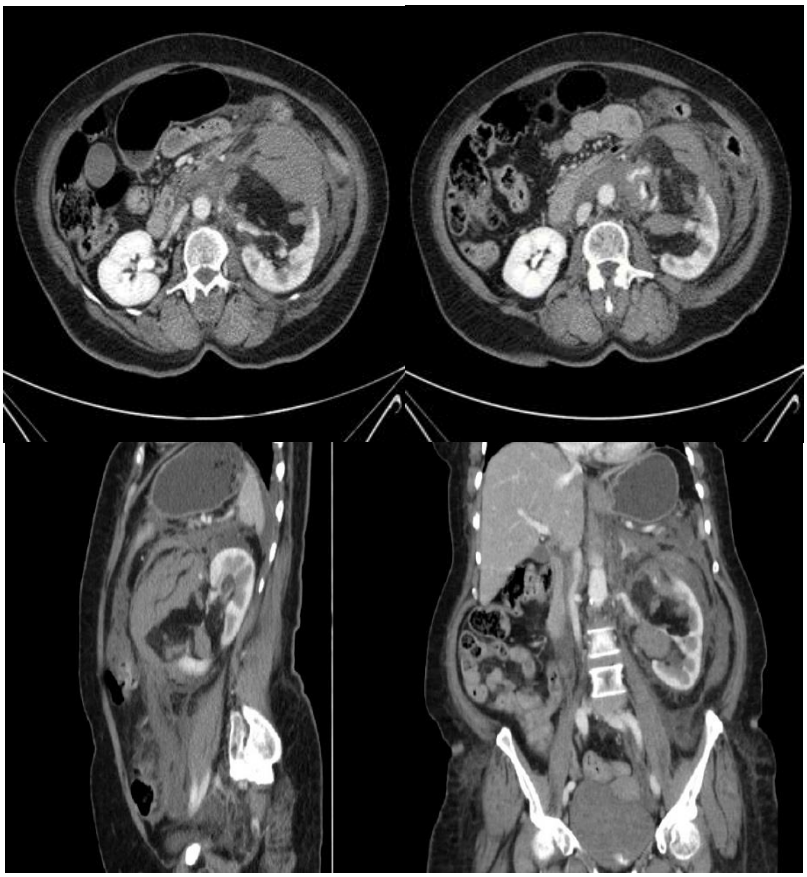

Figure 1. Abdominal and pelvic CT scan with IV contrast (transverse, sagittal, and coronal views). A soft tissue mass consists of fat tissue and blood vessels in the left kidney, mild right perinephric fluid, and adipose tissue inflammation in the left perineal space. Free fluid around the retroperitoneal vessels and the left colon down to the pelvic cavity were reported.

with deep breathing and cough, did not radiate to any part of the body, and was not reduced by any painkiller. The patient had no history of trauma; however, there was a history of diabetes and hypertension treated with aspirin, metformin, glibenclamide, losartan, and amlodipine. Her initial blood pressure and pulse rates were 90/65 mmHg and 118 b/min, respectively. Her body temperature and respiratory rate were normal. The patient's initial hemoglobin was 11.7 g/dL with a white blood cell count, creatinine, and blood sugar of 11400/µL, 0.9 mg/dL, and 236mg/dL, respectively. Nonetheless, her hemoglobin dropped by 2.3 units in the first 24 h.

After IV fluid resuscitation and stabilization, an abdominal and pelvic ultrasound scan was performed which identified mild hydronephrosis in the left kidney with a mass-like lesion measuring 84×90 mm that occupied the upper zone and pelvis of the left kidney. No calcification or vessel flow was observed in the mass. Hyperechoic regions within the mass probably indicated fat tissue suggestive of angiomyolipoma. Increased mesenteric fat echogenicity was observed around the left kidney, and left perinephric, paracolic, as well as pelvic fluid collections, were observed in this case. The longitudinal diameters of the right and left kidneys were 110 and 124 mm, respectively. The liver, gallbladder, spleen, and pancreas were normal. Abdominal and pelvic CT scan with IV contrast revealed a soft tissue mass consists of fat tissue and blood vessels in the left kidney. The mild right perinephric fluid was also observed, and the adipose tissue inflammation was reported in the left perineal space. Free fluid was noted around the retroperitoneal vessels and the left colon down to the pelvic cavity (Figure 1).

The patient was diagnosed with bleeding due to angiomyolipoma of the left kidney and referred to the surgical ward. She underwent a radical left nephrectomy due to the diagnosis of angiomyolipoma with active bleeding, followed by decreased hemoglobin and worsening of vital signs. The mass was sent for histological studies, and the diagnosis was confirmed in the histological examination. The patient was discharged in a good general condition after a few days of hospitalization.

Discussion

Angiomyolipoma is a benign tumor consisting of adipose tissue, smooth muscle, and blood vessels. This tumor can be sporadic or associated with tuberous sclerosis. Sporadic cases are within the range of 80-90% and are usually observed in 40-70-year-old females. The tumors in these cases are unilateral and focal; however, in cases associated with tuberous sclerosis, they are bilateral and multifocal and can occur regardless of age and gender. Most patients are asymptomatic, and the tumor is often diagnosed accidentally during an ultrasound or CT scan (9, 11, 12). Common clinical symptoms include flank pain, throbbing mass, and hematuria. The main complication of angiomyolipoma is bleeding, which is related to the size of the tumor. Increased vascularity and abnormal thickness of the vessel wall predispose the tumor to microaneurysms and bleeding (13). Wunderlich syndrome is a rare syndrome that occurs spontaneously with subcapsular and perirenal space hemorrhage without a history of trauma (14).

In addition to pain, the patient also had urinary and systemic symptoms and was hemodynamically stable upon admission to the emergency department. However, the patients with larger angiomyolipoma tumors and even worse hemorrhagic status presented with only pain or milder symptoms, compared to the patient in our study (15-18). Angiomyolipoma is rarely observed in pregnant females, which can complicate and even terminate the pregnancy(18). Therefore, the size of the tumor or even the presence of an aneurysm or pseudoaneurysm cannot determine the extent of the patient's symptoms. This condition can also cause disorders in laboratory markers, such as leukocytosis and decreased hemoglobin. Similar cases of bleeding of this tumor have been reported in Iran, and the patients' clinical condition was much better than that of our reported case at the time of referral (16). One of the interesting cases of angiomyolipoma in Iran was the appearance of angiomyolipoma after a kidney transplant (17).

Most cases of angiomyolipomas appear as hyperechoic mass containing fat and soft tissue in ultrasound studies. However, the hyperechoic findings are not specific for angiomyolipomas and can be found in other kidney tumors, including renal cell carcinoma, liposarcoma, and atypical Wilms tumor. The CT scans are very specific and show adipose tissue in the lesion, which confirms the diagnosis (8, 19). A magnetic resonance imaging along with a fat suppression technique can detect fat tissue inside the tumor. Angiographic findings include saccular micro and macro aneurysms, hypervascularity, and tortoiseshell arteries. The absence of adipose tissue in the tumor, surrounding soft tissue involvement, and inferior vena cava invasion can rarely be noted in angiomyolipoma, which makes it difficult to differentiate from renal cell carcinoma (20). However, there have been rare reports of renal cell carcinoma containing fat tissue. Calcification has been observed in these cases (21), wheras angiomyolipoma rarely contains calcification (22). It may be difficult to differentiate large angiomyolipoma from liposarcoma since both of them contain fat. The evaluation of renal parenchyma defects and the presence of large arteries can help differentiate angiomyolipoma from liposarcoma (23). Angiomyolipoma tumors with pain, hematuria, suspected malignancy, large tumor size, and spontaneous rupture should undergo surgery. In sporadic angiomyolipoma, follow-up with ultrasound or CT scan should be considered if the tumor size is less than 4 cm (24). However, embolization angiography may be performed in cases with mild bleeding and stable hemodynamics (1).

Conclusion

Angiomyolipoma can be presented with sub-capsular and perirenal hemorrhage with no history of trauma that can be life-threatening

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Conflict of interest

The authors declare that there is no conflict of interest

References

1. Nelson CP, Sanda MG. Contemporary diagnosis and management of renal angiomyolipoma. J Urol 2002;168(4 Pt 1):1315-25. PMID: 12352384
2. Fittschen A, Wendlik I, Oeztuerk S, Kratzer W, Akinli AS, Haenle MM, et al. Prevalence of sporadic renal angiomyolipoma: a retrospective analysis of 61,389 in-and out-patients. Abdom Imaging 2014;39(5):1009-13. PMID: 24705668
3. Flum AS, Hamoui N, Said MA, Yang XJ, Casalino DD, McGuire BB, et al. Update on the diagnosis and management of renal angiomyolipoma. J Urol 2016;195(4 Pt 1):834-46. PMID: 26612197
4. Katabathina VS, Katre R, Prasad SR, Surabhi VR, Shanbhogue AK, Sunnapwar A. Wunderlich syndrome: cross-sectional imaging review. J Computer Assist Tomogr 2011;35(4):425-33. PMID: 21765296
5. Simmons J, Hussain S, Riley P, Wallace D. Management of renal angiomyolipoma in patients with tuberous sclerosis complex. Oncol Rep 2003;10(1):237-41. PMID: 12469175
6. Kiefer RM, Stavropoulos SW. The role of interventional radiology techniques in the management of renal

angiomyolipomas. Curr Urol Rep 2017;18(5):36. PMID: 28299630
7. Buj Pradilla MJ, Martí Ballesté T, Torra R, Villacampa Aubá F. Recommendations for imaging-based diagnosis and management of renal angiomyolipoma associated with tuberous sclerosis complex. Clin Kidney J 2017;10(6):728-37. PMID: 29225800
8. Woo S, Suh CH, Cho JY, Kim SY, Kim SH. Diagnostic performance of CT for diagnosis of fat-poor angiomyolipoma in patients with renal masses: a systematic review and meta-analysis. AJR Am J Roentgenol 2017;209(5):W297-307. PMID: 28834444
9. Wright TJ, Colon PJ, Vakar-Lopez F, Bakthavatsalam R, Psutka SP. Sporadic Angiomyolipoma of the kidney with associated renal vein tumor thrombus. Curr Problems Cancer 2020;1:100016. Link
10. Majdoub A, Khallouk A, Farih MH. Aggressive renal angiomyolipoma extending to the renal vein: about a case and literature review. Pan Afr Med J 2017;28:190. PMID: 29599888
11. Wang L, Ni D, Zhong L, Wang J. Familial genetic tuberous sclerosis complex associated with bilateral giant renal angiomyolipoma: a case report. Oncol Lett 2017;14(6):7099-106. PMID: 29344140
12. Lee G, Nabavizadeh R, Osunkoya AO, Master VA. A rare case of vena cava tumor thrombus associated with epithelioid angiomyolipoma. Urology 2020;142:e4-7. PMID: 32445769
13. Nason GJ, Morris J, Bhatt JR, Richard PO, Martin L, Ajib K, et al. Natural history of renal angiomyolipoma favors surveillance as an initial approach. Eur Urol Focus 2020;S2405(20):30162. PMID: 32636160
14. Kim JW, Kim JY, Ahn ST, Park TY, Oh MM, Moon DG, et al. Spontaneous perirenal hemorrhage (Wunderlich syndrome): an analysis of 28 cases. Am J Emerg Med 2019;37(1):45-7. PMID: 29779678
15. Wright T, Sooriakumaran P. Renal angiomyolipoma presenting with massive retroperitoneal haemorrhage due to deranged clotting factors: a case report. Cases J 2008; 1(1):213. PMID: 18834543
16. Barghi MR, Rahmani M, Haghighatkhah HR. Large intratumoral pseudoaneurysm in a patient with bleeding renal angiomyolipoma. Iran J Radiol 2006;3(3):189-92. Link
17. Roozbeh J, Eshraghian A, Geramizadeh B, Nikeghbalian S, Salehipour M, Malek-Hosseini SA. A rare incidence of angiomyolipoma after kidney transplantation. Iran J Kidney Dis 2012;6(4):311-3. PMID: 22797103
18. Morales JP, Georganas M, Khan MS, Dasgupta P, Reidy JF. Embolization of a bleeding renal angiomyolipoma in pregnancy: case report and review. Cardiovasc Intervent Radiol 2005;28(2):265-8. PMID: 15696353
19. Qiu T, Ling W, Lu Q, Lu C, Luo Y. Contrast-enhanced ultrasound in diagnosis of epithelioid renal angiomyolipoma with renal vein and inferior vena cava extension. J Med Ultrason 2016;43(3):427-30. PMID: 26960715
20. Hadley D, Bryant L, Ruckle H. Conservative treatment of renal angiomyolipomas in patients with tuberous sclerosis. Clin Nephrol 2006;65(1):22-7. PMID: 16429838
21. Yousaf A, Nabi U, Hussein ML, Twair A, Ben Gashir M. Fat-containing renal cell carcinoma mimicking angiomyolipoma: a radiological and histopathological diagnostic challenge. Cureus 2020;12(1):e6721. PMID: 32133249
22. Cholet C, Eiss D, Cohen D, Verkarre V, Hélénon O. Calcified renal Angiomyolipoma: a case report. Urology 2016;97:e7-8. PMID: 27443470
23. Israel GM, Bosniak MA, Slywotzky CM, Rosen RJ. CT differentiation of large exophytic renal angiomyolipomas and perirenal liposarcomas. Am J Roentgenol 2002;179(3): 769-73. PMID: 12185060
24. Faddegon S, So A. Treatment of angiomyolipoma at a tertiary care centre: the decision between surgery and angioembolization. Can Urol Assoc J 2011;5(6):E138-41. PMID: 21388589