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Angiomyolipoma, Report of a Large Cystic Case for 20 Years

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Abstract Angiomyolipomas (AMLs) are the most common benign tumors of kidneys. Due to the presence of challenges in radiology in some cases, a biopsy is needed for differentiation between benign and malignant tumors and to avoid unnecessary treatment. The patient was a 53-year-old female who presented with a right renal cyst and left flank fullness. The history of the renal cyst was 20 years ago. Ultrasound examination showed a solid-cystic hypervascular mass in favor of cystic renal cell carcinoma or multilocular cystic nephroma. After surgery, the pathologist reported angiomyolipoma with extension to the capsule and tumor greatest diameter of 15cm. Physicians must be aware of this entity, different presentations, radiology, and histopathology aspects, to avoid misdiagnosis and unnecessary treatment.

Keywords: *Angiomyolipoma, case report, kidney neoplasms, cystic degeneration*

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1. Introduction

Angiomyolipomas (AMLs) are the most common benign tumors of kidneys. These tumors are composed of different amounts of vessels, smooth muscle fibers, and fatty tissue as its name indicates. Most cases have enough fat to be diagnosed easily in the radiologic examination, however, in the case of small fat component, radiologic diagnosis is difficult and renal cell carcinoma (RCC) should be ruled out [1]. This is more prominent with little fat and the presence of calcification, which highly suggests malignancy [2]. In cases with a high amount of fatty tissue, well-differentiated liposarcoma is in differential diagnosis [3]. AMLs have prevalence of 0.2% - 0.6% with strong female predominance. Most cases are sporadic. About 20 percent are associated with tuberous sclerosis or lymphangioleiomyomatosis. Along with typical (triphasic) cases in pathology, atypical (monophasic or epithelioid) cases are on record. Due to the presence of challenge in radiology in later cases, a biopsy is needed for differentiation between benign and malignant tumors and to avoid unnecessary treatment [1]. Here we report a large cystic case of AML based on histopathology diagnosis for a better understanding of the disease.

2. Case Report

The patient was a 53-year-old female who presented on February 18th, 2017 with a right renal cyst and left flank fullness. The history of the renal cyst was 20 years ago. The patient was evaluated at that time for lumbar disc hernia and MRI was done. A suspicious lesion made the physician for using ultrasound and CT scan of the kidney. A cystic lesion of the kidney was noticed. Two months ago flank pain made her for medical treatment seeking. No history of dysuria, hematuria, or frequency was present. Past medical history was unremarkable except for thyroid disturbance since 1-2 years ago with hyperthyroidism, euthyroidism, and recently (since 6-7 months ago) hypothyroidism. Drug history was levothyroxine 1/2 tablet daily. Physical exam showed enlargement of the thyroid (goiter) in head and neck exam. Ultrasound examination on December 15th, 2016 showed: At least 3 echogenous well-demarcated masses with a maximum diameter of 16mm in the right liver lobe. Right kidney (103mm) with the cortical thickness of 15mm, a solid-cystic hypervascular mass measuring 140x80x10mm in the lower pole, and right kidney pelvis in favor of cystic renal cell carcinoma or multilocular cystic nephroma was noticed. CT scan of abdomen and pelvis with and without contrast on December 28th, 2016 showed: In posterior segments of the right liver lobe at least two hypodense foci in the subhepatic region each measuring 10-12mm suggestive of hemangioma. Hypodense and multicystic lesion in the middle and lower third of right kidney measuring 90x100mm with internal septa and enhancement, lobulated margin with extension to renal sinuses (multicystic nephroma or other multicystic kidney masses). Lab data was a complete blood count with hemoglobin 9.6 g/dl, WBC =10.9x10^3 /mm^3. Na, K, Urea,

and Cr were normal. The patient underwent surgery on February 19th, 2017 with a clinical impression of right 12x14cm solid cystic renal mass. The specimen consisted of kidney tissue measured 15x7x3cm, attached cyst measured 15x10x7cm, and attached fatty tissue measured 11x6x3cm. Cut sections revealed a multiloculated cystic appearance containing clear fluid (Figure 1). Distance between mass and kidney margin was 1cm. The pathologist reported: compatible with angiomyolipoma (Figure 2 A-D) with extension to the capsule, tumor greatest diameter of 15cm, and unremarkable margin. IHC with Desmin, HMB-45, and CD117 for confirmation was recommended. Written informed consent was obtained from the patient. No further follow-up was available for the pathologist.

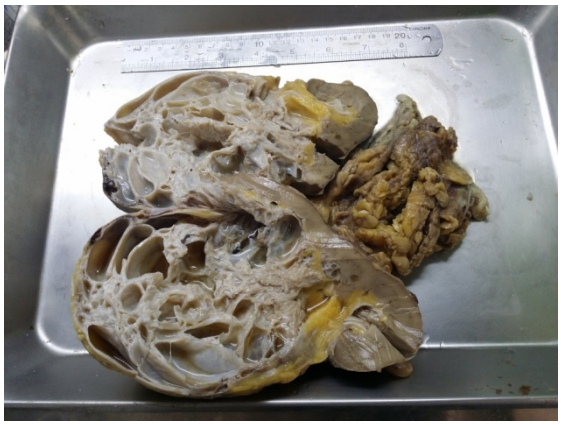

Figure 1. Gross appearance of AML, multicystic renal tissue

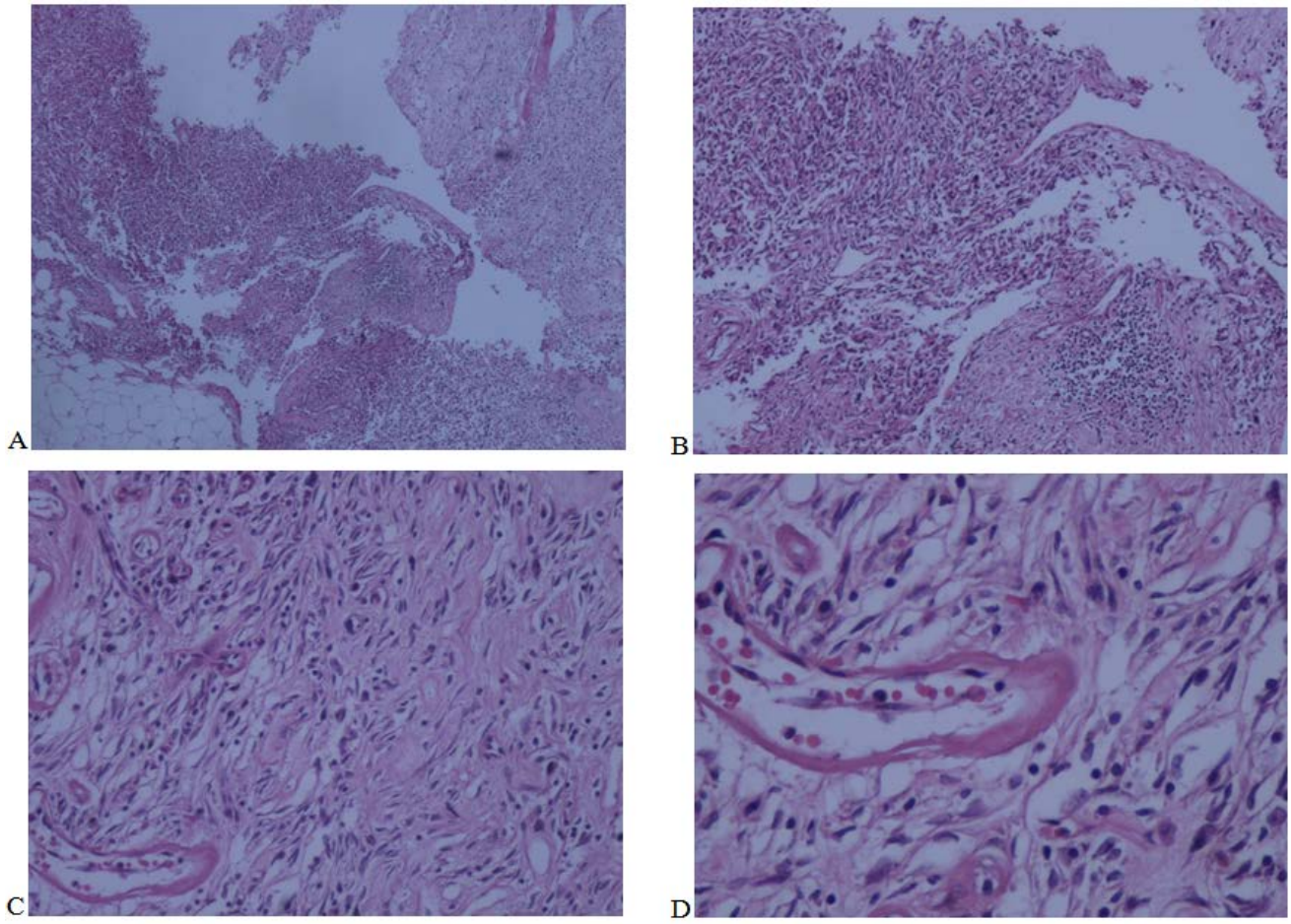

Figure 2. Histopathology of AML, Three components of vessel, smooth muscle fibers, and fatty tissue. Hematoxylin-Eosin staining, X40, X100, X200, X400 magnification (A-D respectively)

3. Discussion

Although AML is the most common tumor in the kidney, it is a rare tumor in urology daily practice [4]. Cases larger than 7 and 20 centimeters are considered as large and giant respectively according to the literature [4,5]. The present case, with a diameter of 15cm, is considered a large AML. Giant tumors as large as 39cm are on record [5]. These tumors are more prevalent in tuberous sclerosis [5]. Guru P Painuly reported a giant AML in an Indian female. The weight of the mass was 1350 grams with a maximum diameter of 30cm of the whole specimen [6]. It is recommended that AML be followed up for at least 5 years after surgery, by imaging and laboratory methods, to rule out any possibility of tumor recurrence [7]. The present case has a history of 20 years with no treatment. Classic renal AMLs with extrarenal extension and vascular invasion are on record [8]. AML is the most common renal tumor with hemorrhage. This event is more probable in large tumors and is a mimicker of ruptured adrenal tumors including pheochromocytoma [9]. Risk factors of hemorrhage are larger tumor size, genetic abnormality, aneurysm, and pregnancy [10]. Necrosis is also present in some cases [3]. The present case with cystic appearance, made the radiologist suggest diagnoses as, multicystic nephroma or multicystic renal tumor. Fat-

containing RCC is a challenge in radiology and pathology that mimics AML. Calcification in the fat-containing tumor is in favor of RCC [11]. Macroscopic fat in a solid renal tumor does not mean AML. In radiologic evaluation increased vascularity, large size, irregular border, and focal calcification are clues in favor of RCC [12]. In histopathology, immunohistochemistry may be helpful. Immunohistochemistry shows HMB45 positivity (100%), MART1/Melan-A, muscle-specific actin (HHF35, 100%), calponin (100%), and NKI-C3 (70–100%). Also, CD117, Desmin, HMB50, microphthalmia transcription factor, Smooth muscle actin, tyrosinase, and vimentin positivity are on record with different percentages. Progesterone receptor is positive in 28% of cases mainly in young females with tuberous sclerosis. Lymphatic differentiation is shown by D2-40 positivity in some cases and albeit S100 positivity in the fat component. HMB45 and Melan-A tend to be diffuse and strongly positive in descending order in fat, smooth muscle, and blood vessels [3]. The pathologist suggested an immunohistochemistry panel of Desmin, HMB-45, and CD117 for this case.

4. Conclusion

Large cases of AML, especially with cystic appearance may be a diagnostic challenge in radiology and histopathology. Physicians must be aware of this entity, different presentations, radiology, and histopathology aspects, to avoid misdiagnosis and unnecessary treatment.

Acknowledgments

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Conflict of Interest

The authors declare that there is no conflict of interest.

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