



Renal Angiomyolipoma Associated with Renal Vein and Inferior Vena Cava Thrombosis: A Case Report

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Abstract

Angiomyolipoma (AML) is the most common benign renal mesenchymal neoplasm. This is a report of a 36-year-old female patient with AML with the involvement of the inferior vena cava (IVC) who was admitted to our hospital. The patient complained of mild right flank pain. CT scan results showed a hypo-dense mass with 47 × 72 mm dimensions at the right kidney's lower pole suggesting renal AML. In MRI with contrast, venous thrombosis was detected in the right renal vein and IVC. Right radical nephrectomy and IVC thrombectomy were successfully conducted. Renal AML was confirmed by pathological findings, and the presence of tumor thrombosis was approved in the right renal vein and IVC. Although AML is generally benign and vascular invasion is a rare finding in this condition, imaging studies (including CT scans) should always be considered to determine the extent of vascular involvement and choose an appropriate therapeutic plan, including nephrectomy and thrombectomy in case of vascular involvement. Despite its benign nature, it should be considered that AML can invade venous structures in the kidneys. Early imaging studies and therapeutic interventions are necessary for obtaining the best outcome.

Keywords: Angiomyolipoma, Flank pain, Inferior vena cava thrombosis, Kidney

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Introduction

Angiomyolipoma (AML) is the most common benign renal mesenchymal neoplasm originating from perivascular epithelioid cells. The tumors comprise a variable proportion of dystrophic vessels, smooth muscle, and adipose tissue (1).

Studies estimate a prevalence of 0.13% for AML in the general population, with a higher proportion in females and a peak incidence in the fourth and fifth decades of life. These tumors can occur sporadically or as a part of genetic syndromes, most commonly tuberous sclerosis complex (TSC) and lymphangioleiomyomatosis (2).

AML is often diagnosed incidentally upon performing routine scans by different imaging techniques particularly CT scans which can confidently detect it (3).

AML may be either asymptomatic or progress to a complicated condition, typically upon a tumor growth beyond 40 mm or the occurrence of a hemorrhage (4).

Although venous involvement is a common complication of renal cell carcinoma (RCC), it rarely occurs in AML because of its benign nature (5). The inferior vena cava (IVC) involvement by renal AML is

particularly an uncommon phenomenon. So far, from only 91 reported cases of venous involvement in AML, 77 have been detected in the IVC (6). We here reported a case of AML with IVC thrombosis.

Case Report

The patient was a 36-year-old woman without any past medical history, complaining of mild pain in the right flank. Clinical examinations and laboratory findings were normal. Abdominal and pelvic sonography revealed a hyper-echo mass with dimensions of 51 × 84 mm at the right kidney's lower pole. CT scan of the same areas also showed a hypo-dense mass of 47 × 72 mm at the lower pole in the lateral margin of the right kidney (Hounsfield unit value: -62), which its vessels were evident after injecting the contrast material. These findings suggested renal AML. Due to suspicion of IVC involvement in the CT scan result, MRI was performed (Figure 1).

In abdominal and pelvic MRI, a mass was seen in the lower-middle pole of the right kidney. The mass enhancement was increased after injecting the contrast material. Along with this, venous tumor thrombosis was



detected in the right renal vein and IVC (level 3: extending to intrahepatic IVC and below the hepatic vein). The patient, with the diagnosis of invasive AML, underwent right radical nephrectomy and IVC thrombectomy by an urologic oncologist and vascular surgeon.

Renal AML was confirmed by histopathologic examination and IHC staining for the melanocytic marker (with cytoplasmic positivity for HMB45 in tumoral epithelioid cells), and the calcified thrombosis was approved in the IVC, and extensive tumoral tissue surrounded it (Figures 2-4).

Discussion

Renal AMLs are benign tumors found sporadically (80%) or in association with TSC (7). Contrary to other benign

renal masses, AML can be diagnosed with renal imaging. The presence of macroscopic fat on a CT scan or MRI is a diagnostic marker for AML. Ultrasonography is not a reliable method in this condition since masses, such as some RCC tumors, are hyperechoic. In a CT scan, the presence of intralesional fat (Hounsfield unit < -15 to -20) on unenhanced series is a diagnostic marker (2).

Rarely, AML may reveal a potentially aggressive behavior such as invading the renal vein, IVC, or regional lymph nodes. Nevertheless, AMLs with malignant features are extremely unusual (1). Only 77 cases of AML with IVC invasion have been published so far (6). The large size and the position of tumors (either center or right) are factors contributing to the invasion of renal veins (1,3,8). Treatment depends on tumor size, the presence or absence of symptoms, and reproductive age, and should be tailored to preserve renal function (2).

To choose among therapeutic options, staging an AML venous thrombus is primordial. Understanding the extent of the thrombus in the IVC lumen is necessary for staging; according to the TNM classification, a T3b



Figure 1. A hypo-dense mass of 47 × 72 mm at the lower pole in the lateral margin of the right kidney in CT scan (AML) (A), vessels were evident after injecting the contrast material in CT scan (B), tumor thrombosis in IVC in MRI (T2) (C, D).



Figure 2. The cut surface of renal tumor after nephrectomy sent for histopathologic evaluations

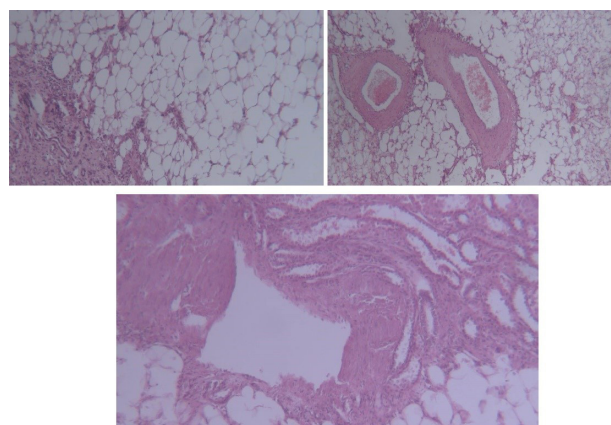


Figure 3. Microscopically (H&E, ×100 magnification) histological findings of three components of AML (admixture of adipose tissue, blood vessels, and smooth muscle tissue)

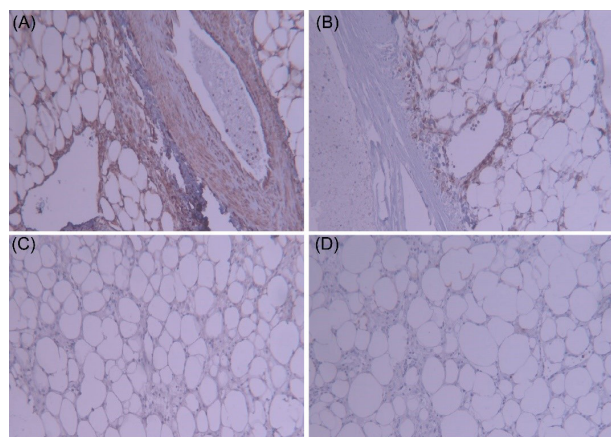


Figure 4. Immunohistochemistry staining shows positive for Vimentin (A) and HMB-45 markers (B) in polygonal epithelioid cells, and negative reaction for CD-10 (C) and Cytokeratin (D) (magnification: ×100)

tumor is below the diaphragm while a T3c tumor is above the diaphragm (9). Nonetheless, it should be always kept in mind that the presence of a thrombus in the renal vein or IVC, regardless of the type of tumor, is an indication of complete nephrectomy and thrombectomy (10). Our patient, according to imaging findings and after being diagnosed with AML, was subjected to right radical nephrectomy and IVC thrombectomy. In pathological findings, AML in the right kidney, thrombus in the renal vein, and IVC were confirmed. Although AML is generally a benign condition, and vascular invasion is an uncommon finding in this condition, it should always be considered that necessary imaging studies (including CT scan) are indicated to perceive the presence and extent of vascular involvement and to choose an appropriate therapeutic plan. Nevertheless, nephrectomy and thrombectomy are indicated in the case of vascular involvement.

Conclusion

Despite its benign nature, it should be considered that AML can invade venous structures. Early imaging studies and therapeutic interventions are necessary for obtaining the best outcome.

Author Contributions

Conceptualization: Reza Mohammadi, Ali Kamalati.

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Validation: Mohammadali Bagherinasabsarab.

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Supervision: Reza Mohammadi, Ali Kamalati.

Conflict of Interests

None.

Ethical Approval

Informed consent was obtained from the patient for the publication of this report.

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