Bilateral Renal Angiomyolipoma with Bilateral Sarcomatous Change

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Abstract

A 54-year-old man presented to the nephrology clinic with three months history of hematuria, weight loss and loss of appetite. Radiological and pathological investigations pointed to the diagnosis of multiple bilateral renal angiomyolipomas with bilateral high grade sarcomatuos transformation and lung metastasis, and with no manifestations of tuberous sclerosis. This is the first reported case of bilateral renal angiomyolipomas with multiple sarcomatous changes.

Keywords: renal angiomyolipoma, sarcomatous transformation, computed tomography.

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Introduction

Typical renal angiomyolipoma (RAM) is a well-characterized benign renal neoplasm in which mesenchymal proliferation composed of thick-walled blood vessels, smooth muscle, and adipose tissue with the occurrence of variable proportions. It is seen either sporadically or as a part of tuberous sclerosis complex where it is considered the most common renal lesion. [1] Sarcomatous transformation is a rare event but was described in the literature in some cases occurring unilaterally. This report presents a case with bilateral sarcomatous transformation in RAM with multiple pulmonary metastases.

Case Report

A 54-year-old man who previously had hypertension and hyperlipidemia, but who is not known to have tuberous sclerosis or a family history of tuberous sclerosis, presented to the nephrology clinic complaining of gross hematuria (clots in the urine), loss of appetite and weight loss of three months duration. His urinalysis showed numerous RBCs, WBCs and +2 proteins. Hemoglobin was 10 g/dl and kidney function tests showed renal impairment (createnine 2.2 mg/dl, urea 8,8 mg/dl).

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He was given a trial of oral antibiotics but he showed no improvement. Two weeks later, he was admitted to our hospital for further evaluation. Renal CT scan completed by a dynamic study revealed multiple bilateral renal masses showing areas of lipid density and patchy heterogenous enhancement consistent radiologically with bilateral RAM. No abnormalities were otherwise detected in the abdominal organs (Fig. 1, 2). Chest CT scan demonstrated the presence of innumerable bilateral lung nodules with an average diameter of 2-5 mm seen predominantly in both lung bases with peripheral location typical radiologically for metastatic lung disease. It is well recognized that lung secondary deposits from sarcomatous primaries may show cavitations and be responsible for pneumothorax ; however, these features were not present in our case (Fig. 3). Brain CT scan revealed no abnormalities.

In view of all findings, renal biopsies were done and the histopathology report demonstrated the presence of pleomorphic cells with pleomorphic hyperchromatic nuclei and frequent abnormal mitotic figures consistent with high grade pleomorphic sarcoma (Fig. 4, 5). Bilateral nephrectomy was planned and discussed with the patient who refused any intervention and died at home few months later.

Fig. 2: Reconstructed coronal CT scan image of both kidneys illustrating the renal masses.

Fig. 3: Axial chest CT scan (lung window) showing multiple bilateral lung nodules due to metastasis.

Fig. 1: Axial CT scan image with I.V contrast showing multiple bilateral heterogeneous enhancing masses causing renal enlargement and distortion.

Fig. 4: Section shows a core needle biopsy of tumor tissue that consists of pleomorphic cells with pleomorphic hyperchromatic nuclei with ample eosinophilic cytoplasm. (20x)

Fig. 5: The section shows a closer view of the tumor cells with bizarre giant nuclei and frequent abnormal mitotic figures. (40x)

Discussion

RAM was previously regarded as a hamartomatous lesion characterized by proliferation of thick-walled blood vessels, smooth muscle cells and fat in variable proportions, however it is now considered a neoplasm arising from a common progenitor cell that can proliferate and differentiate to produce each of the three classic tissue components of angiomyolipoma called the perivascular epithelioid cell. [1] It is the most common benign solid renal tumour recognized on modern imaging with an estimated prevalence of 0.3-3%. [3]

RAM occur either sporadically or as a part of tuberous sclerosis complex. In both sporadic renal tumours and in patients with clinical manifestations or a family history of tuberous sclerosis, the presence of a renal mass with fat density on radiologic imaging permits a presumptive diagnosis of angiomyolipoma. [3] It is a slow growing tumour and although typical angiomyolipoma is considered a benign tumour, it is thought that the epithelioid variant of angiomyolipoma may behave in an aggressive manner. [2] Rarely, angiomyolipoma may display malignant potential with rapid growth and metastasis, the malignancies include malignant epithelioid angiomyolipoma [5-7] and sarcomatous transformation of one of the tumour components; most commonly leiomyosarcoma. [8]

It is agreed now that cellular atypia, venous extension, intra/extra renal multifocality and local recurrence after incomplete surgical resection should not be interpreted as malignant. [2,9-12] The diagnosis of sarcoma arising from angiomyolipoma requires metastasis and fatal outcome for acceptance as true sarcomatous transformation. [13] Malignant transformation is considered rare and was only reported in both sporadic cases and those associated with tuberous sclerosis in a few cases in the literature, and they were all unilateral.

This report presents a patient who did not have any other manifestations of tuberous sclerosis (supported by normal clinical examination and brain CT scan) nor had a family member with the disease. Radiological investigations showed multiple bilateral heterogeneously enhancing renal masses demonstrating areas of fat density and multiple pulmonary nodules consistent with metastasis, these findings together with the pathology reports led to the diagnosis of bilateral sarcomatous changes in multiple renal angiomyolipomas with pulmonary metastatic disease and fatal outcome. Although the molecular testing for tuberous sclerosis was not done, the disease was excluded on clinical and radiological grounds and on absence of family history.

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تحولات غرنية في الورم الوعائي العضلي الدهني في كلتا الكليتين

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الملخص

نعرض اليكم فيما يلي الحالة المرضية التالية التي تم تشخيصها في مستشفى الجامعة الأردنية:

مريض عمره (54) سنة كان يعاني من ارتفاع التوتر الشرياني وارتفاع في مستوى الدهنيات في الدم، وكان المريض موضوعاً على العلاج الدوائي لارتفاع الضغط الدموي الشرياني، تم فحص المريض في عيادة امراض الكلى حيث كان يعاني من بيلة دموية ونقص شهية وانخفاض واضح في وزن الجسم لعدة أشهر قبل قدومه الى المستشفى.

تم استقصاء المريض سريرياً ومخبرياً وشعاعياً حيث شخص المريض أعلاه ورم وعائي عضلي دهني في كلتا الكليتين (أورام متعددة) وبالفحص النسيجي والتشريح المرضي تبين أن هنالك تحولات نسيجية غرنية سرطانية (عالية الخباثة) وكان لدى المريض نقائل ورمية للرئتين، وقد توفي المريض بعد بضعة أشهر لاحقة للتشخيص، علماً بأن المريض لم يشخص عنده تصلب أو في أي من أفراد العائلة.

ومن تدقيق الحالات النادرة للتحولات الغرنية في الورم العضلي الوعائي الدهني في الكلية هو احادي الجانب بينما ما تتميز به الحالة المرضية التي تعرضها لكم هو وجود ورم ثنائي الجانب.

الكلمات الدالة: الورم الوعائي العضلي الدهني الكلوي، التحولات الغرنية، التصوير الطبقي المحوري.