A rapidly progressive type 2 papillary renal carcinoma in a 63-year-old kidney donor: case report

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Abstract

Introduction. Papillary renal carcinoma is a heterogenous disease and the second most common histological type of renal malignant tumor after clear cell carcinoma. Of the three types, type 2 papillary renal carcinoma has the worst impact on patient heath status.
Materials and method. We present the case of a 63-year-old man, former kidney donor with type two papillary renal carcinoma with rapid growth and fast disease progression to metastatic stage. Although upon been investigated for kidney donation and being declared disease free and fit for the kidney transplantation, this patient developed 12 years later a fast growing renal neoplasia.
Conclusions. Because type 2 papillary carcinoma has a worse prognosis than other renal malignancies, with rapid progression from local to metastatic stage, the treatment of these cases should not be postponed and patient should be rapidly assessed by urologist and if necessary by oncologist. Kidney transplantation remains the best solution for people with end-stage renal failure but we must also keep in mind that the donor which is left with solitary kidney must be reevaluated periodically because it has a higher risk that general population of further renal pathology on the urogenital apparatus.

Key words: papillary renal carcinoma, kidney transplantation, renal cancer

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Introduction

Papillary renal carcinoma is the second most common histological type of renal malignant tumor. According to EAU Guidelines 2015, papillary RCC is a heterogenous disease, that has three different subtypes: 1, 2 and oncocytic. The prognosis of subtype 2 papillary carcinoma is worse than subtype 1 (1).

Type 1 papillary renal carcinoma can be found in a familial syndrome of renal carcinoma more frequent than type 2 papillary renal carcinoma (3).

Papillary renal carcinoma is a multi-focal disease, that has a peak of incidence in the 6th and 7th decades of life, and it is more frequent in the male population rather than in the female population (2).

Case presentation

A 63-year-old male presented to our clinic with an ultrasonography performed in a clinic in his home town that suggested the presence of a possible tumor in the right kidney. The patient was a former kidney donor - he had donated his left kidney to his son 12 years prior.

He was admitted in our clinic on the 30th of June 2014. A contrast enhanced CT scan of the abdomen and pelvis was performed. The CT findings were (fig. 1- fig. 4): big, encapsulated tumor with diameters of 13/9/12 cm located in the left lumbar area with possible origin the adrenal gland; a 2.1/2.4 cm tumor located in the upper pole of the right kidney; a small mass (0.7 cm), with apparently no malignancy criteria located in the right adrenal gland; no adenopathies or other metastasis were found on the CT in the abdomino-pelvic region.

The findings are resumed in the images below:

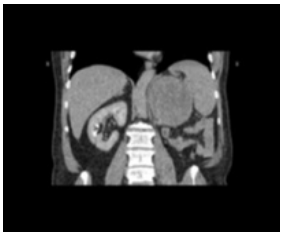

Fig 1: left adrenal gland tumor

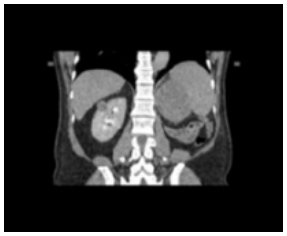

Fig 2: left adrenal gland tumor and right kidney tumor

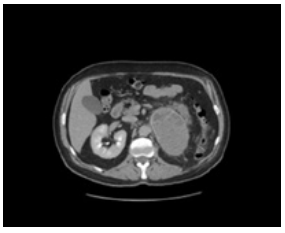

Fig 3: left adrenal gland tumor

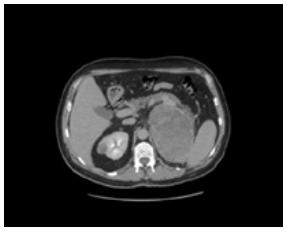

Fig 4: left adrenal gland tumor and right kidney tumor

The patient was scheduled for surgery on the 10th of July 2014 and a partial nephrectomy via lumbar approach was performed. There were no periprocedural complications and the blood loss was minimal.

On the 2nd day postoperatively the patient presented dyspnea, polypnea, with O_2 Sat of 50%, PaO_2 of 36 mmHG, blood pressure of 150/80 mmHg, heart rate of 160 bpm. An emergency contrast enhanced CT scan of thorax was performed that diagnosed massive bilateral pulmonary thromboembolism. Specific treatment was immediately initiated with heparin and oxygen therapy.

On the 20th of July 2014 (10th day postoperatively) another contrast enhanced CT scan of thorax was performed to reassess the lungs; it revealed an important regression of bilateral thromboembolism. The patient was switched to oral anticoagulant. There were no other complications so the patient was discharged on the 2nd of august and was instructed to come back to the hospital 6 weeks later for reevaluation and therapeutic decision regarding the tumor in the left adrenal gland.

The pathological report revealed that the tumor in the right kidney was a type 2 renal papillary carcinoma, pT1, Fuhrman grade 3.

As instructed the patient returned to the hospital and was readmitted on the 25th of September 2014 for further investigations and treatment regarding the tumor in the left adrenal gland.

A contrast enhanced CT scan was performed to reassess the lesion in the adrenal gland and it revealed an increase in size for the tumor in the left adrenal gland – at that time it measured 17/10.8/15.3 cm - and 2 new lesions that had appeared in the right adrenal gland, the biggest of them measuring 2.6/2.6 cm. The imaging diagnosis was secondary malignant lesions.

The CT findings are illustrated in the images below (Fig. 5-8):

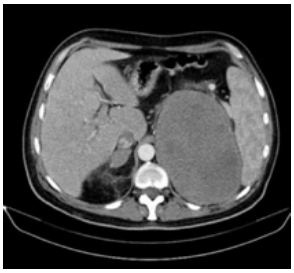

Fig 5: tumor in right adrenal gland

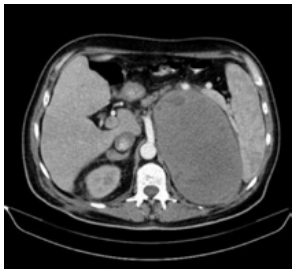

Fig 6: left and right adrenal gland tumors

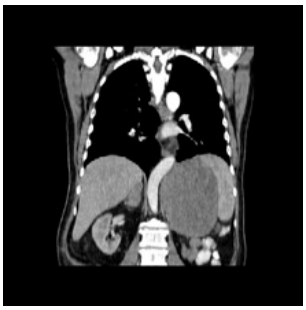

Fig 7: left adrenal gland tumor

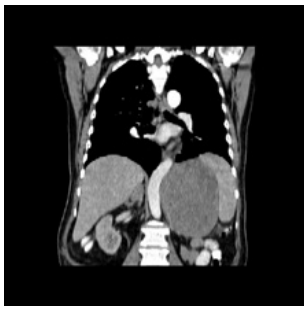

Fig 8: left and right adrenal gland tumor

The oncologic status of the patient was reassessed and it was decided that the patient should undergo a left adrenalectomy, which was performed transperitoneally on the 6th of October 2014. During the surgery, because of local tumor spread, it was necessary to perform a splenectomy. There were no other periprocedural complications and the blood loss was minimal. The patient recovered better and faster after this second surgery.

The pathological report from the surgery was consistent with metastasis from renal papillary type 2 carcinoma.

The patient was discharged after one week of hospitalization. Because of splenectomy and because of the massive bilateral pulmonary thromboembolism that had occurred under correct anticoagulant therapy we decided to refer the patient to the hematology department for further investigations and management of the concomitant hematological status.

He was admitted in the hematology department in December 2014. The bloodwork done revealed a modified hemoleucogram, consistent with the splenectomy that we had to perform on the second surgery. Nonetheless a bone marrow biopsy was indicated and performed by the hematologist. The pathological report form the bone marrow biopsy indicated the presence of malignant cells in the bone marrow that were consistent with metastasis from the papillary renal tumor.

Upon receiving the biopsy results a new contrast enhanced CT scan was performed that showed the presence of the two lesions in the left adrenal gland, osteocondensant lesions in the pelvic region (on the left side of the iliac bone and pubis) and left inguinal adenopathy (that measured 4/2.5 cm).

The lesions are shown in the images below (Fig. 9-11):

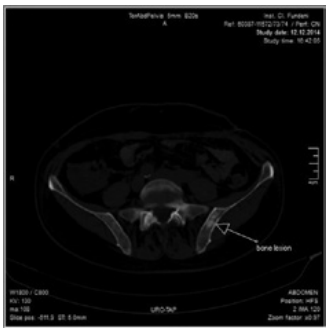

Fig 9: bone lesion in left iliac bone

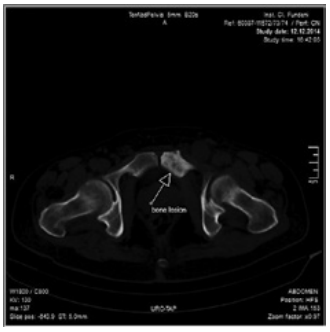

Fig 10: bone lesion left branch of pubic bone

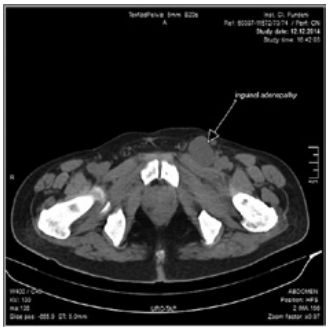

Fig 11: left inguinal adenopathy

A bone scan was also performed that showed multiple sites suggestive for bone metastasis, located in the rib cage, right humerus, pelvic bones, both femurs and tibiae. The patient was considered beyond surgical resources and was referred to the oncology department for chemotherapy.

Discussions

According to literature review type 2 papillary renal carcinoma has a worse prognosis that type 1 papillary carcinoma (4) and is quicker to progress to metastatic stage. We must keep in mind that type 1 and 2 papillary renal carcinoma are different in prognosis and management than the newly described oncocytc variant – or type 3 - that has a very low malignant potential (8). Also we must underline the fact that no high volume studies have been reported on this particular type of renal cancer (4,5) thus making it a challenge for the urologist in both therapeutic and follow-up management.

We must underline that both disease free survival and overall survival are considerably lower in type 2 papillary carcinoma than type 1 papillary carcinoma, according to literature review. According to one study that followed 130 patients with papillary renal carcinoma for a time period of up to 111 months, overall and disease-free survival rate was 89% and 92% in type 1 papillary renal carcinoma and 55% and 44% in type 2 papillary renal carcinoma (5).

The particularity of this case is that the patient is a former kidney donor that was fully investigated in 2003 when he donated his left kidney to his son. He had no suspect lesions at that time and was considered eligible for the operation. 12 years later he became an oncologic patient with a rare type of renal carcinoma.

Furthermore we must underline the fast growth of the tumor masses and rapid disease progression to a metastatic stage even if according to literature patients with papillary renal carcinoma are less likely to present with T3 or greater disease (11). Also the lymphovascular invasion is less important in type 2 papillary renal carcinoma than in type 1 or in clear cell renal carcinoma (13). Nevertheless there are studies that have shown an important association between type 2 papillary renal carcinoma and vena cava thrombus (12), but fortunately this was not the case here.

We must also keep in mind that his son, the recipient of the renal graft is, at this moment, in perfect clinical and imagistic condition, even if he receives immunosuppression therapy for his kidney transplant. Even if the chance for developing a tumor after renal transplantation is low (9,10) – according to some studies in the literature it is lower than 5%, the recipient is now in a very strict active surveillance follow-up program in order to have any abnormalities quickly discovered in regard to his renal graft.

Conclusions

Because type 2 papillary carcinoma has a worse prognosis than other renal malignancies, with rapid progression from local to metastatic stage, the treatment of these cases should not be postponed and patient should be rapidly assessed by urologist and if necessary by oncologist.

Kidney transplantation remains the best solution for people with end-stage renal failure but we must also keep in mind that the donor which is left with solitary kidney must be reevaluated periodically because it has a higher risk that general population of further renal pathology on the urogenital apparatus.

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