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Case Report

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Unusual Synchronous Ipsilateral Different Histological Patterns in the Same Kidney: Impact of Genetic Mutation on Tumor Aggressiveness and Complexity

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Abstract

Renal cell carcinoma (RCC) is a rare visceral neoplasm and the presence of epididymis-like tubules is even rarer, where only 26 cases have been reported in the literature. The presence of synchronous different histologic types of RCC in the same kidney is extremely rare in clinical practice. Minimally invasive radical and partial nephrectomy represent the cornerstone in the management of renal tumors. This report describes a patient with left renal mass undergoing laparoscopic radical nephrectomy, where histopathology identified four different histological types, consisting of a multifocal papillary RCC in the middle and lower pole, oncocytoma in upper pole, multifocal papillary adenoma, and epididymal-like tubules, together with a review of the current literature.

Keywords: Renal cell carcinoma, Histopathology, oncocytoma, papillary adenoma, Epididymis-like Tubules

Introduction

Being the most common primary renal neoplasm, Renal Cell Carcinoma (RCC) is a rare malignancy, which account for 1-3% of all visceral neoplasms. RCC comprises different histological subtypes and few cases of ipsilateral multiple histological lesions have been described, including most commonly a mixture of clear cell with chromophobe or papillary RCC [1]. Imaging studies may help suspecting the presence of tumors of different histologic subtypes within the same kidney. However, most cases are only identified by histopathological examination after surgical excision, which represent the definitive management for RCC. This case report will present a patient undergoing laparoscopic radical nephrectomy for a left renal mass consisted of four different histopathological patterns in the same kidney, including multifocal papillary RCC and adenomas, oncocytoma, and epididymal-like tubules.

Case Presentation

A 77-years-old man referred with a left renal mass, which was incidentally discovered during the work up for a previously resected right leg Schwannoma. The patient was asymptomatic with no abdominal complaint, hematuria, or bother some voiding symptoms. Physical examination revealed jaundice, bruising, skin discoloration and skin rashes. The patient had a normal preoperative renal profile and complete blood picture. Contrast-enhanced Computed Tomography (CT) showed bilateral multiple cortical cysts suggestive of mild form of polycystic disease, the largest measured 4.0 x 4.2 x 3.9 cm in the left kidney. The right kidney cysts showed fluid density while there were three enhanced lesions on the left kidney, including a 2-cm soft tissue density lesion in the upper pole, a 2.6 cm soft tissue density in the lower pole, and amid-pole exophytic lesion, which demonstrated intermediate density with moderate peripheral heterogeneous enhancement. There were no associated lymph node enlargements, venous involvement, or pelvic or abdominal metastasis **(Figure 1)**.

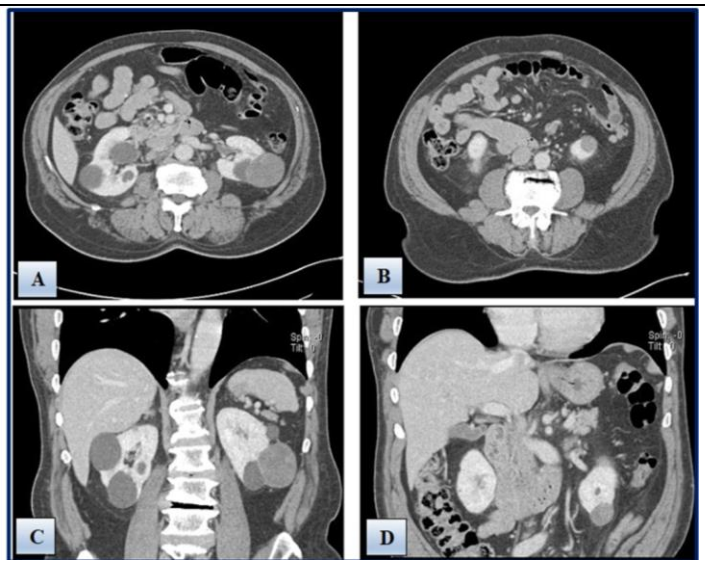

Figure1: Axial (A & B) and Coronal (C & D) sections of CT abdomen with contrast showed bilateral multiple renal cysts suggestive of mild form of polycystic disease. The right cysts are fluid density while on the left side three lesions are enhancing. A 20 mm enhancing soft tissue density at the upper pole, a 47 mm mid pole lesion is exophytic mass, demonstrating intermediate density with moderate peripheral heterogeneous enhancement, and a 26 mm soft tissue mass in the inferior pole.

The patient underwent a laparoscopic left radical nephrectomy, with no perioperative adverse events. Histopathological findings identified pT1a multifocal RCC in the middle and lower poles, the largest ones were 4.0 x 3.5 x 3.5 cm and 2.0 x 2.0 x 2.0 cm, with Fuhrman nuclear grade 2. A multifocal papillary adenoma was also noted, with an upper pole oncocytoma measuring 1.5 cm in its greatest dimension. Epididymal like tubules were seen in renal sinus, as well **(Figure 2)**. Next-generation sequencing, genetic analysis of kidney cancer showed heterozygous variant c. 995G>A p (Arg332His) in

the PBRM1 gene. The patient has a smooth postoperative course with stable vital signs and discharged home in a good general condition with normal renal profile and blood count. Two-years postoperatively, the patients had no evidence of local recurrence or distant metastasis with a normal serum creatinine.

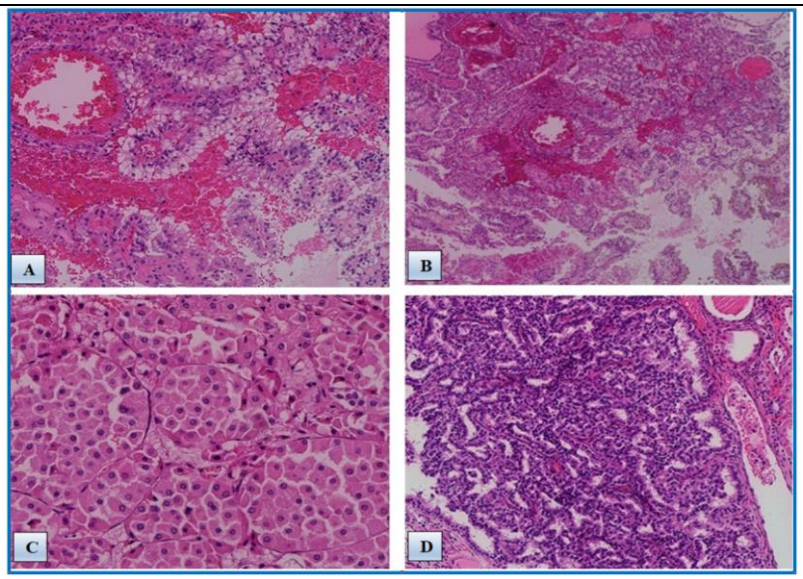

Figure 2: Histopathological examination showedpapillary RCC having clear cytoplasm (A), Neoplasm with papillary architecture (B), Oncocytoma with cells which are large with abundant cytoplasm (C), Papillary adenoma with tubulopapillary architecture (D).

Discussion

RCC consists of different morphological sub types, where clear cell type represents the most common form of by 70%, followed by papillary RCC by 10-15% of all RCCs [2]. Considerable morphologic heterogeneity within a single tumor significantly preclude capturing the aggressive behavior of the entire tumor, making the situation much harder with ipsilateral different histological patterns. RCC with different Ipsilateral renal lesions have been previously reported [3,4]. The most common reported mixed pattern was a mixture of clear cell with chromophobe or papillary RCC [1]. The present indexed case is unique for the different benign and malignant histological patterns, including multifocal RCC and adenomas, oncocytoma, and the epididymis-like tubules. Despite that histopathological and immunohistochemical staining revealed T1a RCC, recurrences are still expected in 50% of patients, even after 5-year follow-up. Patients with similar multiple Ipsilateral renal tumors have a 5-fold possibility of contra lateral kidney recurrence, what necessitate longer thorough follow-up time than those with solitary renal tumors [6]. Our patient has completed 2-year follow-up with contrast enhanced CT scans without evidence of local recurrence or distant metastasis. The presence of epididymis-like tubules in a renal specimen is even rarer and

previously reported only in 26 cases of renal dysplasia; most of them were case reports [5]. These tubules are mostly of mesonephric rather than metanephric origin due to their branching and communication with the calyces, together with absence of associated glomeruli [5]. The authors suggested that associated aberrant expression of some markers might be due to the dysplastic kidneys. Of interest, most peripheral nerve tumors, including neurofibromas and schwannomas, are benign and a rise from the nerves heath with the two most common being. Schwannoma has been reported in metastatic RCC via spread from the primary mass rather than the rare hematogenous seeding to a peripheral nerve [7]. Our patient has a history of a previously resected leg Schwannoma, whose follow-up workup was the main reason for the incidentally discovered renal masses indexed in the current case. Three to five percent of RCC are related to mutations a susceptibility gene, which increases the risk of developing specific RCC subtypes. Recent advances in Next-Generation Sequencing (NGS) have led to explore these genes and to the identification of additional new susceptibility genes. In the present case, tumor multimodality and personal history guide referral for cancer geneticist, where we have explored a panel of genes in germline. The genetic test result revealed a variant of unknown significance in PBRM1 gene. In somatic genomic landscape, the PBRM1 gene is mutated in approximately 40-50% of patients. Loss of PBRM1 function in tumor suppression has been correlated with advanced tumor and more aggressive features, such as poor differentiation and lymphovascular invasion [8], and poor prognosis, in terms of recurrence free and overall survival [9]. The detected variant in PBRM1 gene may play a role in our patient tumor complexity more studies (somatic and functional) are needed. The patient underwent minimally invasive laparoscopic radical rather than partial nephrectomy, considering that the challenging multifocal tumors with unpredictable pathology. Histologic detection of multifocal RCC in the middle and lower renal poles supported the need for more aggressive therapy. This surgical approach was a safe and effective treatment option for such a complex case with rare synchronous four different histological patterns in the same kidney, with two-year recurrence-free status, the last follow-up time.

Conclusion

This report described a complex case with rare synchronous four different histological patterns in the same kidney. PBRM1 gene may play a role in tumor aggressiveness and complexity. Minimally invasive laparoscopic nephrectomy was a safe and effective treatment option with recurrence-free status or distant metastasis until the last follow-up time of 2-year postoperatively.

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