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Mucinous Tubular and Spindle Cell Renal Cell Carcinoma: Case Report and Literature Review

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ABSTRACT

Mucinous tubular and spindle cell renal cell carcinoma is a rare tumor described in the 2004 WHO classification as a new entity with an indolent clinical course. In this study, we report the case of a 60-year-old man with a large mass involving the upper pole of the left kidney. The patient underwent left radical nephrectomy. The histological findings showed it to be a mucinous tubular and spindle cell renal cell carcinoma of grade 1 of Fuhrman's classification with hilar infiltration. Because of the favorable outcome of this histological entity, mucinous tubular and spindle cell carcinoma must be differentiated from papillary renal cell carcinoma, especially from the variant with sarcomatoid dedifferentiation. The aim of this work is to draw the attention of pathologists and clinicians to this new entity, and the importance of its diagnosis for the patient's prognosis.

Introduction

Mucinous tubular and spindle cell renal cell carcinoma (MTSRCC) is a rare renal cell carcinoma (RCC) described for the first time in 1998. It was previously included in the category of unclassified renal cell carcinoma (RCC) in the World Health Organization (WHO) tumor classification [1]. In 2004, it has been recognized as a single entity: a variant of RCC [2]. Although it has been reported that MTSRCC has a relatively good prognosis, the data remain limited in order to establish its clinical behavior. Thus, we present the case of a low grade MTSRCC in a 60-year-old man in order to enrich the literature with a new case, and we will proceed to a literature review.

Patient and observation

A 60-year-old smoker (30 pack-years) male patient, with no medical history, consulted for a mild left lumbar pain for 2 months, with a single episode of haematuria, without any other neither urinary nor digestive symptoms. His physical examination was unremarkable: no fever, no anemic syndrome, and no palpable lymph nodes.

The computed tomography (CT) scan shows a well limited, hypodense, slightly enhanced after contrast injection, large tissular mass of 20 cm, involving the upper pole of the left kidney (**FIGURE 1**). This mass is pushing forward the left renal calyces and pedicle, without evidence of adjacent structures infiltration (**FIGURE 2**). We also note the presence of a 10 mm lateroaortic lymph node. Blood analysis was normal.

Surgical exploration with a left subcostal incision found a large retroperitoneal mass developing from left kidney. The patient underwent a radical nephrectomy. The postoperative course was simple.

On macroscopic examination, the kidney contained a 20 cm, solid, pale yellow mass, with foci of hemorrhage.

This tumor was not well circumscribed, partially encapsulated, and in contact with the renal capsule.

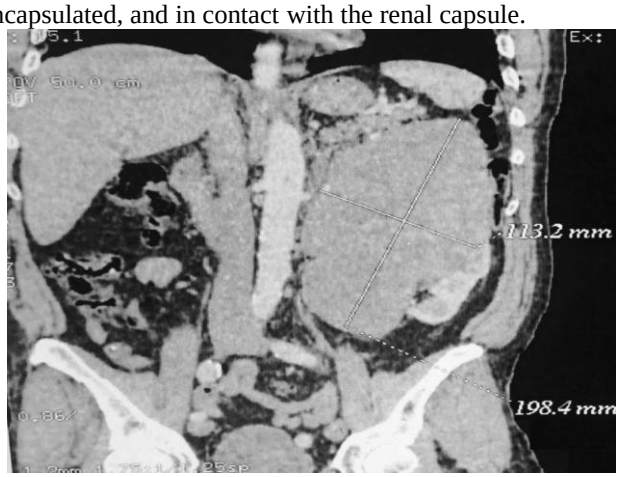

Figure 1. Frontal CT section showing the mass involving the upper pole of the left kidney.

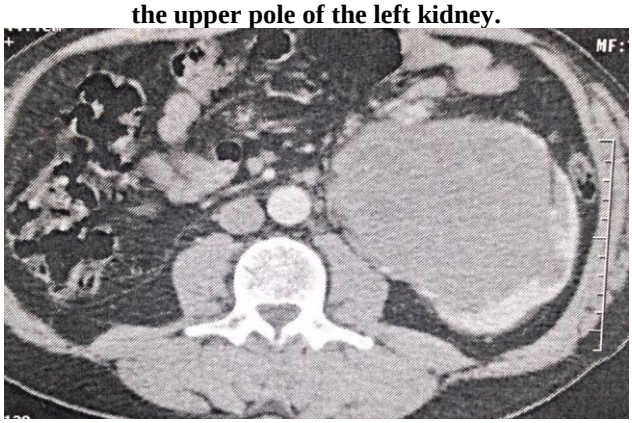

Figure 2. Axial CT section showing the hypodense mass slightly enhanced after contrast injection, occupying the hilar region.

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Histological examination revealed a proliferation of monotonic cubic eosinophilic epithelial cells. Those cells are arranged in tubular structures and parallel cellular cords. These tubular structures are in continuity with fusiform sectors, which are also of low nuclear grades. The microvacuolar stroma contains a mucinous substance with an inflammatory infiltration by histiocytes (**FIGURE 3 and 4**). Neither lymphovascular embols, nor capsular invasion, nor sarcomatoid contingent were seen. This examination concluded to a low grade (grade 1 of Fuhrman) MTSRCC.

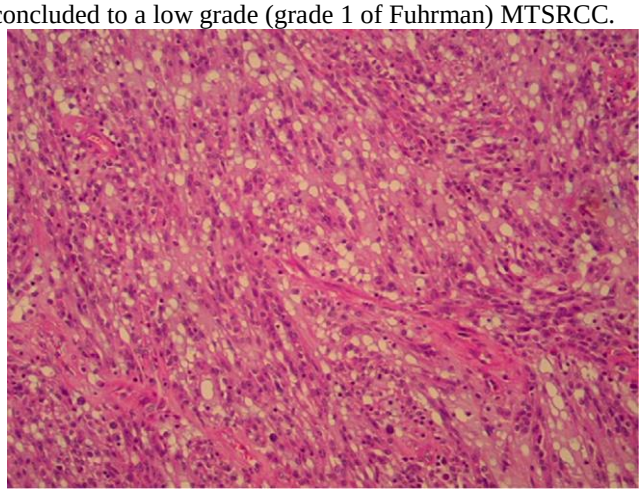

Figure 3. Fusiform cell proliferation of low nuclear grade with an eosinophilic cytoplasm (HES, × 40).

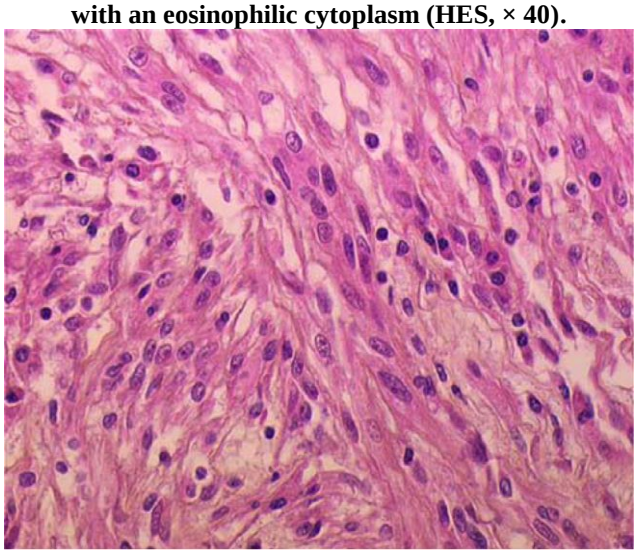

Figure 4. Proliferation of tubular and fusiform architecture in a microvacuolar myxoid stroma (HE, × 250).

Discussion

MTSRCC is a rare malignant epithelial tumor of the kidney, recognized since 2004 as a new entity of RCC. The MTSRCC have the peculiarity of having a feminine predominance and a relatively good prognosis [3]. More than 80 cases have been listed in the literature [2]. However, the rare morphology of the CTM has caused diagnostic problems to many clinicians in the past. The MTSRCC architecture showing focal papillae and a high mitotic activity may be in favor of papillary carcinoma of the kidney. In cases examination find a predominantly fusiform configuration, this may be confused with a leiomyoma or even a sarcoma [4].

Symptomatology is non-specific and can associate haematuria, flank pain and palpable mass [5]. Radiological investigations find calcifications in 34 %, six times higher than in other kidney tumors. There is also a greater frequency of retroperitoneal invasion and a greater tumor hypo-vascularization. However, there are no specific imaging criteria for MTSRCC diagnosis and these characteristics may suggest other variants of RCC, such as chromophobe RCC or papillary RCC, which have a less favorable outcome. Although, the MTSRCC should be suspected in front of a large, well circumscribed mass, slightly enhanced after injection of contrast agent, in association with urolithiasis [1]. The mass size is variable, and ranges from less than 1 cm to more than 18 cm, most tumors measuring 2 to 4 cm [6]. Macroscopically, the epicenter is essentially cortical, well limited, solid, often homogeneous, gray-whitish or more rarely brownish. Areas of necrosis or hemorrhage can rarely be seen [7].

Microscopically, the tumor is characterized by a combination of tubular and spindle cell components separated by a mucinous stroma. The tubules can be round, ovoid, or elongated with a collapsed central lumen. In some cases, the spindle cell component can be dominant, evoking a leiomyoma or myofibroblastoma [8,9,10]. The mucinous stroma is usually abundant, but sometimes can appear as multiple small vacuoles imitating clear cells. The tumor cells have a slightly eosinophilic cytoplasm and indistinctive borders. The nuclei are generally round with low nuclear grade feature, but high nuclear grade can be observed. Mitoses are rare and necrosis is uncommonly seen. Nevertheless, MTSRCC with sarcomatoid differentiation (high nuclear grade, tumor necrosis, and high mitotic activity) have already been reported [11].

These tumors have a complex phenotype, expressing a wide variety of epithelial markers (EMA, AE1 / AE3, CK7, CK19) and distal nephron markers (EMA, CK 19, E-cadherin). The immunohistochemical profiles of these tumors reported in the literature are not clearly defined, they are inconstant and even contradictory because of the heterogeneity of these tumors and the insufficient number of cases studied [8,12,13]. Indeed, the expression of the RCC-Ma is inconstant. A recent study showed that 92 % of the cases studied expressed this marker [4]. In other series, we found an expression in only 45 % and 7 % [13]. Other markers are also variable expression such as EMA, CD 15 and PS 100.

It is evident that tumors having a similar morphological appearance may have different immunohistochemical expression without necessarily implying that they are different. Several classifications of tumors in different organs have been mainly based on their morphology, the immunophenotype being not always exactly the same for tumors classified in the same type. Some authors proposed to classify tumors with the morphological characteristics of MTSRCC according to the expression of different markers [14]. However, the expression of these antigens does not appear to be sufficient to separate tumors with the same clinical presentation, microscopic and evolutionary characteristics.

Genomic investigations for MTSRCC based on karyotyping and fluorescent in situ hybridization (FISH) analyses, showed multiple chromosomal numerical aberrations in these tumors, with losses in chromosomes 1, 4, 6, 8, 9, 11, 13, 14, 15, 18, 22 and X, or gains in chromosomes 2, 3, 4, 5, 7, 9, 10, 12, 15, 16, 17, 18, 19, 20, 22 and Y [11,15].

The prognosis, according to the data of the literature, seems favorable, and corresponds to expectations given the low nuclear grade.

However, cases with local tumor recurrence, distant sites or lymphatic metastases have been reported in the literature [12,16,17,18]. Recently, 5 cases of MTSRCC with sarcomatoid differentiation and aggressive behavior have been reported in the literature. Among these five cases, three had distant metastases with a fatal outcome for patients [17,19]. Therefore, patients with localized stage are must be treated with resection, either partial or radical nephrectomy. For metastatic stages, there are actually no guideline published [20].

Conclusion

MTSRCC is a new entity in the World Health Organization (WHO) tumor classification. It is essential to identify the MTSRCC because of its favorable prognosis. Other cytogenetic and immunohistochemical studies, as well as a greater clinical experience are needed for better characterization of these tumors.

Competing interests

The authors declare no competing interest.

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Author contributions

All authors contributed in the development of this publication and approved the final manuscript.

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