CASE REPORT

UDC: 616.61-006
DOI: https://doi.org/10.2298/VSP190523108L

A case report of an unusual kidney tumor: Mucinous tubular and spindle cell carcinoma

Prikaz bolesnika sa retkim tumorom bubrega – mucinozni tubularni vretenastoćelijski karcinom

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Abstract

Introduction. Mucinous tubular and spindle cell carcinoma of the kidney (MTSCC) is a rare and recently described subtype of renal cell carcinoma (RCC). As its name indicates, the tumor is composed of three morphological structures: tubules consisted of cuboidal cells, spindle cells, and extracellular mucus. **Case report.** A 59-year-old female patient was admitted to the Emergency Center of the Clinical Center of Vojvodina due to injuries sustained in a traffic accident. After diagnostic procedures, computer tomography (CT) revealed a large asymptomatic cyst of the upper pole of the left kidney as an incidental finding. Four months later, after recovering from traumatic injuries, a control CT confirmed a well-circumscribed tumor, 90 mm in diameter, confined to a kidney, and a radical nephrectomy was performed. Histopathological evaluation showed that the necrotic tumor mass consisted of tubules made of cuboidal cells and cords made of spindle cells separated by pale mucinous material in some areas, while other tumor parts were of dense cellularity without mucin. No atypia was found. **Conclusion.** MTSCC is a variant of papillary RCC, thus, it is usually mistaken with papillary RCC with sarcomatoid differentiation. Because of the same immunoprofile as papillary RCC, histomorphology is imposed as the gold standard for making the diagnosis. MTSCC is a tumor with a generally favorable prognosis, and complete surgical excision appears to be adequate treatment, but single cases with metastatic disease have been reported. In this case, there were no signs of the disease one year after surgery.

Key words:
kidney neoplasms; carcinoma, renal cell; adenocarcinoma mucinous; diagnosis; nephrectomy; treatment outcome.

Apstrakt

Uvod. Mucinozni tubularni vretenastoćelijski karcinom bubrega (MTSCC) je redak i nedavno opisan podtip karcinoma bubrežnih ćelija (RCC). Kao što mu ime kaže, tumor se sastoji od tri morfološki različite strukture: tubuli sačinjeni od kubičnih ćelija, zatim vretenaste ćelije i ekstracelularna sluz. **Prikaz bolesnika.** U radu je prikazana 59-godišnja bolesnica koja je primljena u Urgentni centar Kliničkog centra Vojvodine zbog povreda nastalih u saobraćajnoj nesreći. Nakon dijagnostičkih procedura, kompjuterskom tomografijom (CT) otkrivena je asimptomatska velika cista gornjeg pola levog bubrega kao uzgredni nalaz. Četiri meseca kasnije, nakon oporavka od traumatskih povreda, kontrolnim CT pregledom potvrđen je dobro ograničen tumor, promera 90 mm, ograničen na bubreg, zbog čega je izvršena radikalna nefrektomija. Histopatološkom analizom ustanovljeno je da se radi o nekrotičnoj tumorskoj masi sastavljenoj od tubula sagrađenih od kubičnih ćelija i snopova vretenastih ćelija, razdvojenih bledim mucinoznim materijalom u nekim područjima, dok su ostali delovi tumora bili bez produkcije mucina i prisustva atipije. **Zaključak.** MTSCC je varijanta papilarnog RCC, pa se obično pogrešno dijagnostikuje kao papilarni RCC sa sarkomatoidnom diferencijacijom. Zbog istog imunoprofila sa papilarnim RCC, histomorfologija predstavlja zlatni standard za postavljanje dijagnoze. MTSCC je tumor sa, generalno povoljnom prognozom i kompletna hirurška ekscizija je adekvatan tretman lečenja, iako su opisani pojedinačni slučajevi metastatske bolesti. Kod ove bolesnice nije bilo znakova bolesti godinu dana nakon operacije.

Ključne reči:
bubreg, neoplazme; karcinom bubrežnih ćelija; adenokarcinom, mucinozni; dijagnoza; nefrektomija; lečenje, ishod.

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Introduction

Mucinous tubular and spindle cell carcinoma of the kidney (MTSCC) is a rare and recently described subtype of renal cell carcinoma (RCC), which has been recognized as a specific entity in the 2004 World Health Organization (WHO) classification of renal cell carcinoma [1]. To this date, there are approximately 100 cases described in the literature [2]. The tumor mostly affects adults with a mean age of 53 years (range 18–82) with marked female predominance (4:1) as in our case [1], in contrast to RCC. MTSCC is a cytologically low-grade neoplasm, but both high-grade MTSCC and MTSCC with sarcomatoid differentiation are described [3, 4].

Clinical symptoms such as flank pain, abdominal mass, and hematuria are rare but possible [5]; most MTSCC tumors are solitary and were incidentally discovered by ultrasound or computer tomography examinations.

As its name indicates, the tumor is composed of three morphologically different structures: tubules consisted of cuboidal cells, spindle cells, and extracellular mucus and it mostly resembles type 1 papillary RCC but without true papilla formation [6]. In both cuboidal and spindle cells, nuclear atypia and mitoses are rare. The proportion of those morphological parts is different in different cases, and hemorrhage and/or necrosis, minor areas with clear cell and oncocytic change, are possible to find as well [7, 8]. Focal papillations may be found and are usually mistaken with papillary RCC with sarcomatoid differentiation.

Although the literature shows contradictory reports regarding its histogenesis [9, 10], the morphological, immunohistochemical, and genetic features suggest differentiation from collecting duct epithelium. Moreover, immunohistochemical analyses show expression of epithelial membrane antigen (EMA), alpha-methylacyl-coenzyme A racemase (AMACR), cytokeratin 7 (CK7), PAX-8, and vimentin in 80–100% of the cases, which confirms a possible origin from distal convoluted epithelial cells [11]. AMACR expression can also be seen in proximal tubule cells. Therefore, there is a belief that it could be a variant of papillary RCC [10]. CD-10 and RCC are often negative.

MTSCC is a renal tumor with an excellent prognosis, but it is not exclusive because there are single cases with the metastatic disease described.

Case report

In December 2017, a 59-year-old female patient was admitted to the Emergency Center of the Clinical Center of Vojvodina due to injuries sustained in a traffic accident. After diagnostic procedures, a serial fracture of ribs was registered, some subcutaneous hematomas, and no signs of internal bleeding. Moreover, computer tomography (CT) revealed a large cyst on the left kidney as an incidental finding without symptoms (Figure 1A). After recovering from traumatic injuries, the patient came to the Urology Clinic of the Clinical Center of Vojvodina because of hematuria. In April 2018, a control CT was made, and it confirmed the existence of a round, clearly demarcated, inhomogeneous mass, 85 millimeters in size, which changed the kidney contour and made pressure on calyces of the medium part and the upper pole of the kidney (Figure 1B). The lumen of the cyst was filled with debris-like material, and surgical treatment was indicated. A left radical nephrectomy was performed in June 2018, and the specimen was sent to the Pathology Department. Grossly, it was a well-circumscribed tumor, 90 mm in diameter, confined to the kidney with the necrotic content inside. A detailed histopathological examination verified the tumor mass characterized by a mixture of compressed anastomosing and tightly packed and parallelly arranged tubular structures consisted of cuboidal cells with round, pale nuclei (Figure 2A). Atypia was minimal. Tubular structures were separated by thin bundles of spindle cells and variable amounts of extra-

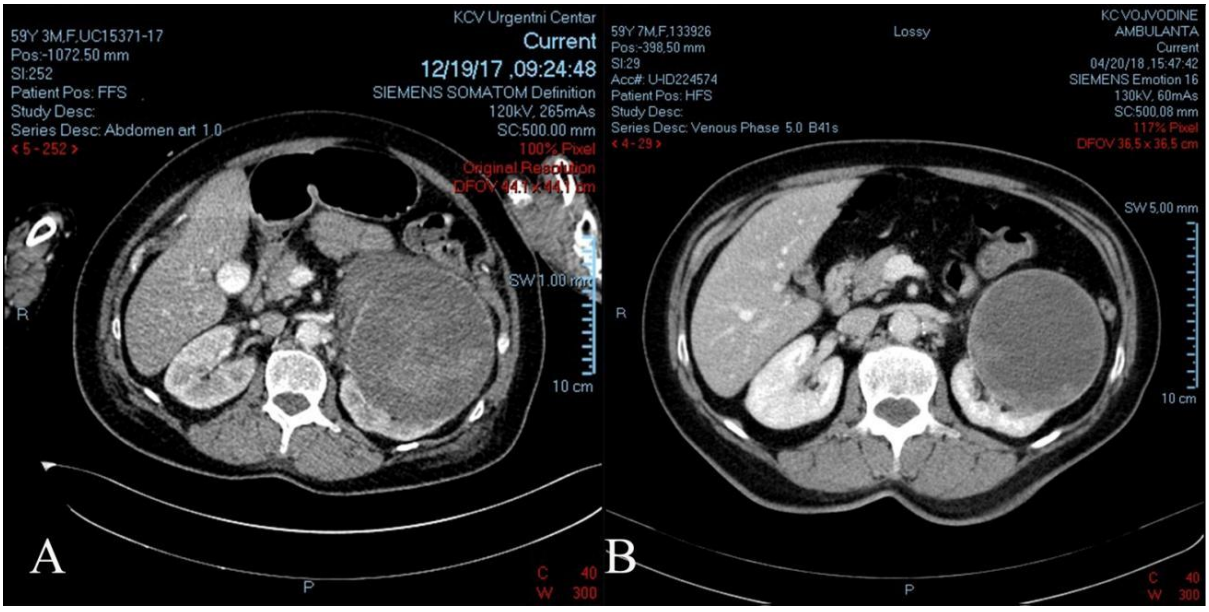

Fig. 1 – A) The first computer tomography (CT) scan showed a big cyst of the left kidney; B) Control CT with the same kidney cyst.

cellular blue-gray Alcian-blue-periodic-acid-Schiff positive mucinous matrix (Figure 2B). Extensive areas of necrosis were present without lymphovascular invasion. Special immunohistochemical stainings showed positivity for CK7 (Figure 2C), AMACR (Figure 2D), Vimentin (Figure 2E), as well as EMA and E-cadherin. RCC and CD-10 were negative. Based on the histological and immunohistochemical description of the tumor, the diagnosis of mucinous tubular and spindle cell renal cell carcinoma was made.

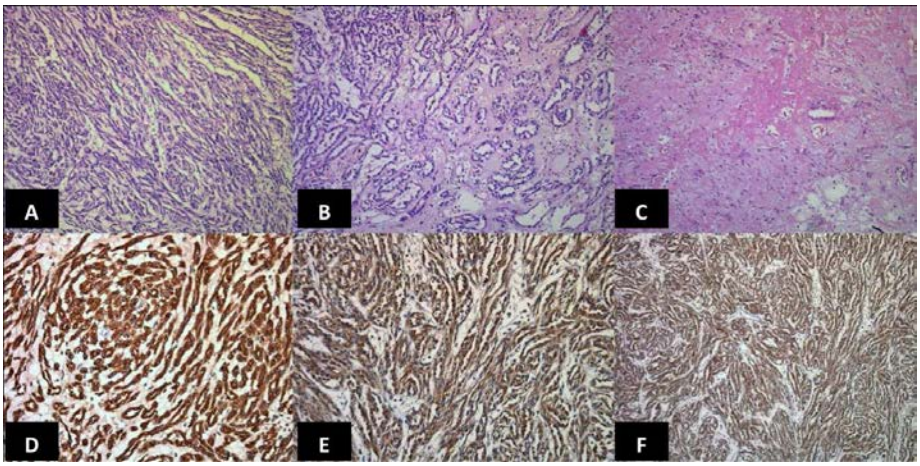

Fig. 2 – A) Spindle cell tumor component [hematohylin and eosin (HE), 10×]; B) Tubular structures of cuboidal cells (HE, 10×); C) Extracellular mucinous matrix (HE, 10×); D) Antibody cytokeratin 7 (CK7) (IHC, 20×); E) Antibody alpha-methylacyl-coenzyme (AMACR) (IHC, 20×); F) Antibody Vimentin (IHC, 10×). IHC – immunohistochemistry.

Discussion

Mucinous tubular and spindle cell renal cell carcinoma is a rare renal tumor. The histogenesis of this tumor is controversial. It has been shown differentiation toward the distal nephron, which led to different tumors, including low-grade tubular mucinous renal neoplasms and low-grade myxoid tumor. Recently, the literature showed immunohistochemical overlap with papillary RCC pointing to proximal tubular differentiation. In fact, MTSCC is a variant of papillary RCC. Although a close relationship to papillary RCC has been suggested, clinically, morphologically, and genetically, it is a distinct renal neoplastic entity. Histopathological examination is imperative for making the correct diagnosis. Immunohistochemistry is not a very useful method for discriminating between papillary RCC and MTSCC, thus histomorphology appears to be the gold standard for making the diagnosis. It is a tumor with a generally favorable prognosis, and complete surgical excision appears to be adequate treatment, but cases with metastatic disease have been reported [12]. Generally, it is estimated that 20–40% of patients with RCC develop metastases after surgery [13]. An unfavorable course of the disease like recurrence, regional lymph nodes, and distant sites metastases, or even death, are associated with atypical histological features such as high nuclear grade and sarcomatoid transformation [14, 15]. Although an innocent outcome is likely, a close follow-up is recommended. In our patient, there were no signs of the disease one year after surgery.

Conclusion

MTSCC is a variant of papillary RCC, hence it is usually mistaken with papillary RCC with sarcomatoid differentiation. Because of the same immunoprofile as papillary RCC, histomorphology is imposed as the gold standard for making the diagnosis. MTSCC is a tumor with a generally favorable prognosis, and complete surgical excision appears to be adequate treatment, but cases with metastatic disease have been reported. In this patient, there were no signs of the disease one year after surgery.

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Received on May 23, 2019.
Accepted September 30, 2019.
Online First October, 2019.