DOI: https://doi.org/10.2298/SARH200617072T
UDC: 616.61-006

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Renal cell carcinoma of a horseshoe kidney

Milomir Tufegdžić[1], Vladimir Vasić[1], Jovan Hadži-Đokić[2]

[1]University Medical Center Zvezdara, Department of Urology, Belgrade, Serbia;
[2]Serbian Academy of Sciences and Arts, Belgrade, Serbia

SUMMARY

Introduction Horseshoe kidney is the most common developmental disorder of the urinary system, which involves an anomaly of kidney fusion, and occurs in 3% of the population. Kidneys are most often connected at the lower poles by fibrous or parenchymal isthmus. Renal cell carcinoma (RCC) is the most common tumor of the horseshoe kidney. Treatment involves surgical treatment that includes heminephrectomy or partial nephrectomy with different approaches. We report a case of RCC of a horseshoe kidney, located on the lower pole and the isthmus.

Case outline A 68-year-old patient reported to the urologist due to intermittent painless macroscopic hematuria. Computed tomography urogram revealed the presence of a tumor on the right kidney, measuring 85 × 90 × 60 mm, with radiological characteristics of RCC, which covered the entire lower pole of the kidney towards the isthmus. Angiography finding indicated a thickened isthmus with pronounced malformation of vascular structures. The right heminephrectomy was performed with resection of the isthmus from 15 mm to the healthy tissue. The isthmus was sutured in two layers with a catgut suture. Subsequently, hilar, paracaval, and interaortocaval lymphadenectomies were performed. The pathohistological finding indicated a tumor of renal cell origin, while the resection line was free of the tumor tissue, as were the lymph nodes.

Conclusion RCC is the most common neoplasm of the horseshoe kidney. Treatment is surgical and involves open or laparoscopic heminephrectomy or partial nephrectomy with a transperitoneal or extraperitoneal approach.

Keywords: horseshoe kidney; renal cell tumor; nephrectomy

INTRODUCTION

Horseshoe kidney is the most common developmental disorder of the urinary system, which involves an anomaly of kidney fusion, and occurs in 3% of the population. This anomaly occurs in the fetal period, between the fourth and sixth weeks of life, and occurs in 1–4 people per 1000 births [1]. Horseshoe kidney occurs more often in men than in the female population in the ratio of 2:1. Developmental disorders of other parts of the urogenital tract as well as other organ systems may be associated with the appearance of the horseshoe kidney. Kidneys are most often connected at the lower poles by the fibrous or the parenchymal isthmus, which can have its own blood vessels [2].

Renal cell carcinoma (RCC) is the most common tumor of the horseshoe kidney, but the risk of developing of this disease is similar to the risk in kidneys without developmental disorders. The highest incidence of RCC is in the seventh and eighth decades of life, while the most common risk factors for RCC are tobacco exposure, obesity, and hypertension. The risk of developing Wilms' tumor and tumors of transitional epithelium increases two to six times in horseshoe kidneys [3]. The diagnosis of the horseshoe kidney is most often made accidentally due to the examination of other diseases and conditions such as arterial hypertension. Symptoms that may indicate the presence of a malignant process are hematuria and nonspecific abdominal pain. Diagnostic procedures include ultrasonographic examination, cystoscopy, angiography, computed tomography (CT) with urography and nuclear magnetic resonance of the abdomen and the lesser pelvis [2, 3].

Treatment involves surgical treatment that includes heminephrectomy or partial nephrectomy with different approaches. These surgical procedures can be complicated by the presence of pathological vascularization and the impossibility of resection of the isthmus. We report a case of RCC of a horseshoe kidney, located on the lower pole and the isthmus.

CASE REPORT

A 68-year-old patient reported to the urologist due to intermittent painless macroscopic hematuria that lasted two months. Complete blood and biochemical analyses were within the reference values. The urine culture was sterile. Arterial hypertension was common comorbidity. Ultrasonographic examination revealed a tumor mass of the right kidney measuring over 80 mm, which extended interpolarly towards the lower pole of the kidney. The lower pole of the kidney was connected to the left kidney by an isthmus. An ultrasound examination indicated a dilatation of the pyelocalyx system of the right kidney, while the pyelocalyx system of the left kidney

Received • Примљено: June 17, 2020
Accepted • Прихваћено: August 30, 2020
Online first: September 15, 2020

Correspondence to:
Vladimir VASIĆ
Marijane Gregoran 15/04
11000 Belgrade, Serbia
vladimirdrvasic@gmail.com

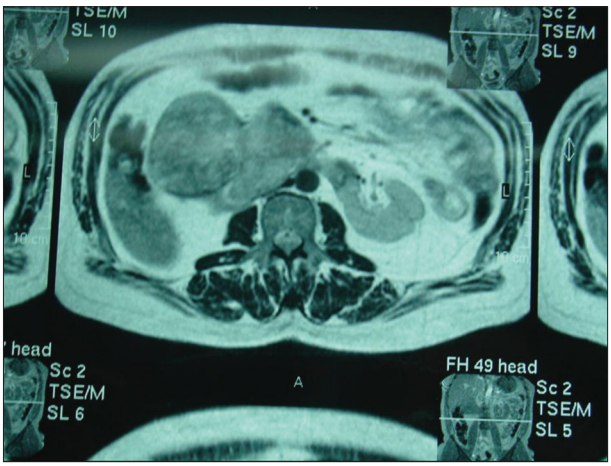

Figure 1. Computed tomography urography showed the presence of a tumor on the right kidney measuring 85 × 90 × 60 mm, with radiological characteristics of renal cell carcinoma, which covered the entire lower pole of the kidney towards the isthmus

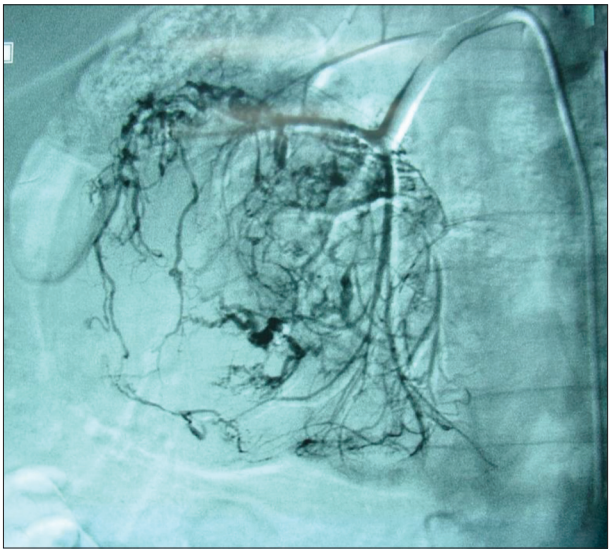

Figure 2. Angiography finding indicated a thickened isthmus with pronounced malformation of vascular structures

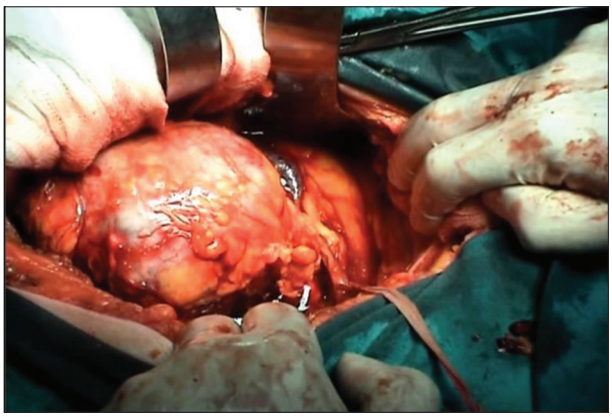

Figure 3. Right heminephrectomy by extraperitoneal approach, with an enlarged lumbotomy incision and resection of rib XI

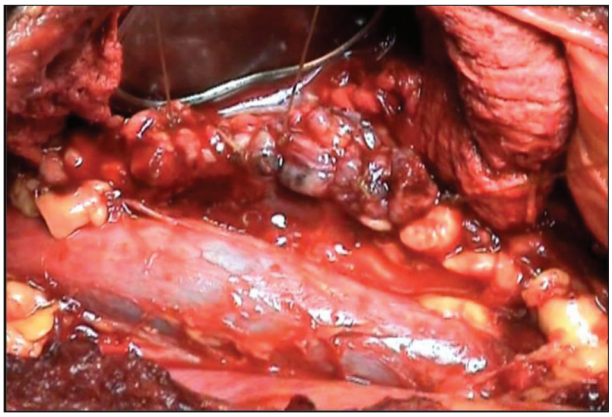

Figure 4. The isthmus was sutured in two layers with a catgut suture; hilar, paracaval, and interaortocaval lymphadenectomies were performed

was normal, without calculosis. Next, CT urography was performed, which showed the presence of a tumor on the right kidney, measuring 85 × 90 × 60 mm, with radiological characteristics of RCC, which covered the entire lower pole of the kidney towards the isthmus (Figure 1). The pyelocalyx system of the right kidney and the initial part of the right ureter were dilated. The left pyelon and the left ureter along the entire length of the course were normal. The angiography finding indicated thickened isthmus with pronounced malformation of vascular structures (Figure 2). It was decided to perform a right heminephrectomy by extraperitoneal approach, with an enlarged lumbotomy incision and resection of rib XI (Figure 3). The right kidney with the tumor mass, the right ureter, the inferior vena cava, and the entire isthmus were dissected and the left ureter was identified. Next, a right heminephrectomy was performed with resection of the isthmus from 15 mm to the healthy tissue. A sample of isthmus tissue from the resection line was sent for pathohistological analysis. The isthmus was sutured in two layers with a catgut suture (Figure 4). The hemostasis was correct. Subsequently, hilar, paracaval, and interaortocaval lymphadenectomy were performed. The postoperative course was uneventful, and the patient was discharged from the hospital on the 10th postoperative day. The pathohistological finding indicated a tumor of renal cell origin, while the resection line was free of the tumor tissue, as were the lymph nodes. After three years, a control CT finding of the abdomen, the pelvis, and the chest showed no local recurrence of the disease or the presence of secondary deposits.

This case report was approved by the institutional ethics committee, and written consent was obtained from the patient for the publication of this case report and any accompanying images.

DISCUSSION

Horseshoe kidney is the most common anomaly of kidney fusion and it is more common in men than in women. Its frequency is estimated at about 0.25% of the total population. Genetic predisposition has not been proven, although this anomaly has been identified in twins and siblings within the same family. The horseshoe kidney consists of two different functional kidneys on each side of the medial

line of the body, which are connected to the lower poles by the isthmus. The isthmus contains blood vessels, functional tissue of the renal parenchyma, or connective tissue [4, 5].

Renal cell tumor is the most common neoplasm that occurs on the horseshoe kidney in about 50% of cases, while transitional cell carcinoma and Wilms' tumor are present in about 25% of cases. The etiological factors are not completely known, but the carcinogenesis of RCC cannot be related to the presence of a congenital defect [6]. Predisposing factors for the development of transitional cell carcinoma of the horseshoe kidney are obstruction, chronic infection and the presence of calculosis. The tumor can be localized in any part of the kidney, but is mostly located on the isthmus [1, 6]. In 1976, David Buntley presented 111 cases of tumors that developed in horseshoe kidneys, where RCC had the highest frequency [1]. Similar data have been published in contemporary literature.

The presence of this anomaly is in most cases asymptomatic and is detected incidentally in the process of diagnosing other processes and conditions. Symptoms are associated with hydronephrosis, infection, and calculus formation. Hematuria and nonspecific abdominal pain may indicate horseshoe kidney malignancy [7]. Abdominal aortic aneurysms and ovarian tumors must be considered as differential diagnoses. Diagnosis of horseshoe kidney tumors involves the same radiological methods used in the diagnosis of physiological development of kidney diseases, which include ultrasonographic examination of the abdomen and the lesser pelvis, CT urogram, angiography and nuclear magnetic resonance of the abdomen and the lesser pelvis [7, 8].

Heminephrectomy is indicated for the localization of tumors in the central part of the kidney, for tumors of larger dimensions, tumors in unfavorable position to the pyelocalyx system and vascular stem, in cases of a thicker and highly vascularized isthmus, as well as in elderly patients. Heminephrectomy usually involves a transperitoneal approach, while in certain cases the method can be reported with an extraperitoneal lumbotomy approach [4, 9].

If the tumor mass is localized at the poles, and its dimensions do not exceed 40 mm, the method of choice is sparing surgery, partial nephrectomy, which involves extraperitoneal and transperitoneal approach, or laparoscopic method using the same approaches [10]. When setting the indication for partial nephrectomy, three important characteristics of the horseshoe kidney must be taken into account: malformations of blood vessels, parenchymal or fibrous structure of the isthmus, and the anatomical position of the kidney itself [11]. In about 30% of cases of horseshoe kidney, one renal artery was observed, while a large number of vascular anomalies are found in about 70% of cases. The structure of the isthmus and the malformations of the blood vessels that supply it bear great influence on the choice of surgical method [12, 13]. In certain cases, preoperative superselective embolization of pathological blood vessels of the tumor can be performed, which enables a more efficient and safer surgical treatment [14].

In the presented case, the tumor measuring over 80 mm affected the entire lower pole and the isthmus, which was extremely thickened with a large number of blood vessels, so it was decided to perform heminephrectomy by the extraperitoneal approach, with an enlarged lumbotomy incision and resection of rib XI.

RCC is the most common neoplasm of the horseshoe kidney. The incidence of this tumor on the horseshoe kidney is not higher in relation to the population with normal fetal kidney formation. Diagnostic methods and prognostic factors are the same as for kidney tumors with undisturbed development. Treatment is surgical and involves open or laparoscopic heminephrectomy or partial nephrectomy with a transperitoneal or extraperitoneal approach. Significant characteristics of the horseshoe kidney that must be taken into account: malformations of blood vessels, parenchymal or fibrous structure of the isthmus, and the anatomical position of the kidney itself, which can determine the type of surgical treatment.

Conflict of interest: None declared.

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Карцином бубрежног паренхима на потковичастом бубрегу

Миломир Туфегџић[1], Владимир Васић[1], Јован Хаџи-Ђокић[2]

[1]Клиничко-болнички центар „Звездара“, Клиничко одељење урологије, Београд, Србија;
[2]Српска академија наука и уметности, Београд, Србија

САЖЕТАК

Увод Потковичаст бубрег је најчешћи развојни поремећај мокраћног система који подразумева аномалију фузије бубрега и јавља се код 3% становништва. Бубрези су преко доњих полова повезани истмусом, који може бити изграђен од везивног или паренхимског ткива бубрега. Карцином бубрежног паренхима најчешћи је тумор на потковичастом бубрегу. Лечење је хируршко и подразумева хеминефректомију или парцијалну нефректомију са различитим приступима. Приказујемо случај карцинома бубрежног паренхима потковичастог бубрега, локализованог на доњем полу и истмусу.

Приказ болесника Шездесетосмогодишњи болесник јавио се урологу због повремене безболне макроскопске хематурије. КТ урографија је показала присуство тумора на десном бубрегу, димензија 85 × 90 × 60 *mm*, са радиолошким карактеристикама карцинома бубрежног паренхима, који је заузимао цео доњи пол бубрега према истмусу. Налаз ангиографије указивао је на задебљани истмус са израженом малформацијом васкуларних структура. Изведена је десна хеминефректомија са ресекцијом истмуса од 15 *mm* до у здраво ткиво. Истмус је ушивен у два слоја са катгутним шавом. После тога су урађене хиларна, паракавална и интераортокавална лимфаденектомија. Патохистолошки налаз указивао је на тумор порекла бубрежних ћелија, док је линија ресекције на истмусу била без туморског ткива, као и лимфни чворови.

Закључак Карцином бубрежног паренхима је најчешћи тумор потковичастог бубрега. Лечење је хируршко и укључује отворену или лапароскопску хеминефректомију или парцијалну нефректомију са трансперитонеалним или екстраперитонеалним приступом.

Кључне речи: потковичаст бубрег; карцином бубрежног паренхима; нефректомија