Upper ureteral calculi complicated with severe hydronephrosis and renal medullary carcinoma in elderly Chinese woman: a case report

Qiwei Yu (✉ dr_yqw1103@163.com)
Kunshan Hospital of Traditional Chinese Medicine

Yufan Wu
Kunshan Hospital of Traditional Chinese Medicine

Linya Yao
Kunshan Hospital of Traditional Chinese Medicine

Xi Zhang
Kunshan Hospital of Traditional Chinese Medicine

Jun Wang
Kunshan Hospital of Traditional Chinese Medicine

Xiang Li
Kunshan Hospital of Traditional Chinese Medicine

Xueming Zeng
Kunshan Hospital of Traditional Chinese Medicine

Case Report

Keywords: Renal medullary carcinoma, Urinary tract stones, Hydronephrosis, Differential diagnosis

Posted Date: September 15th, 2022

DOI: https://doi.org/10.21203/rs.3.rs-2051248/v1

License: ☑️ ☀️ This work is licensed under a Creative Commons Attribution 4.0 International License.
Read Full License
Abstract
Renal medullary carcinoma is a rare and aggressive malignancy that commonly occurs in young African men with sickle cell disease. We report a case of a 64-year-old female patient with left renal medullary carcinoma who was hospitalized for severe hydronephrosis due to upper ureteral calculi obstruction. A laparoscopic left nephrectomy was performed, and the tumor was pathologically diagnosed with renal medullary carcinoma. Postoperative pain was not significantly relieved and the patient developed retroperitoneal metastasis 1 month later and died after 3 months of follow-up. For elderly patients with long-term kidney stones and altered kidney morphology, renal tumors should be considered.

Background
Renal medullary carcinoma (RMC) is an aggressive and very rare malignancy that mostly occurs in adolescents and adult men with sickle cell disease (SCD), mainly of African descent (1–3). The disease was first described in 1995 by Davis(4) and is considered the seventh type of sickle cell nephropathy. Upon RMC diagnosis, patients often present with distant metastases in the lymph nodes, lungs, liver, adrenal glands, and bones. In general, RMC patients have an extremely poor prognosis, with a median overall survival of fewer than 4 months (5). This is primarily driven by the rapid growth of the carcinoma that results in the patient often presenting with gross hematuria, abdominal pain or a palpable mass, and weight loss. Additionally, RMC is occasionally misdiagnosed as a kidney abscess or urinary tract infection, further worsening patient outcomes (6). The current case reports on a 64-year-old female RMC patient who was hospitalized with severe hydronephrosis due to upper ureteral stone obstruction.

Case Report
In March 2021, a 64-year-old woman was hospitalized due to the presence of a left upper quadrant mass and the presence of left low back pain for 4 months. In January, the patient felt that the left low back pain was aggravated, and palpation of the left upper abdomen revealed an obvious mass. The whole abdomen enhanced CT revealed the presence of left ureteral pelvic calculus, left kidney calculus, severe left kidney hydrops, left kidney cortex thinning, and liver cirrhosis. Physical examination revealed a cystic mass approximately 8 cm in diameter in the left upper quadrant with no percussion pain in the bilateral kidneys(Fig. 1). The patient’s laboratory results showed elevated CEA (8.77 ug/L), CA125 (45.33 U/mL), CA199 (256.80 U/mL), and ferritin (301.20 ug/L), and the presence of HbsAg (+).

The patient had a history of hypertension and diabetes, which were treated using long-term medication. In April 2020, the patient underwent percutaneous nephroscopic pneumatic lithotripsy for a "left kidney stone".

The preoperative diagnosis indicated a left non-functioning kidney. After excluding surgical contraindications, a laparoscopic left nephrectomy was performed under general anesthesia on April 1, 2021. During the operation, multiple cysts with an inner diameter of 2–6 cm in the left renal section were
observed. The cyst wall was gray-brown, and necrosis was noted in the cavity. Two nodules measuring 4 x 3 cm and 2 x 2 cm, respectively, were observed in the renal parenchyma surrounding the cyst wall. The cut surface was gray-white-red, with medium texture and an unclear boundary. Postoperative pathology suggested RMC and immunohistochemistry revealed the following indicators: AE1/AE3 (+), Vimentin (+), CD10 (+), CD117 (partially+), TFE3 (weakly+), Mitf (weakly+), PAX-8 (−), PAX-2 (−), INI-1 (−), CAM5.2 (−), EMA (−), CK7 (−), P504S (−), HMB45(−), Melan-A (−), CK20 (−), GATA-3 (−), SDHB (−), and an elevated Ki67 (index 70%) (Fig. 2). The patient was discharged on April 9, 2021. After discharge, the patient's incision did not heal well, and she complained of pain in her left waist. The patient then went to the local hospital for anti-infective symptomatic treatment; however, this treatment did not remedy the pain. On May 17, 2021, the patient went to another hospital for re-examination. There, a whole abdomen enhanced CT showed that there were multiple abnormal density shadows in the left kidney area, left abdominal pelvis, and left abdominal wall. From these data recurrence and metastasis were considered. For economic reasons, the patient did not receive follow-up treatment and died 2 months later.

**Discussion**

RMC is an aggressive and rare malignant tumor. Clinically, 60% of RMC patients have gross hematuria, rib pain, anorexia, weight loss, fever, or the presence of an abdominal mass. Due to the lack of specificity in clinical manifestations, when a patient has urinary tract stones, misdiagnoses may occur that affect subsequent treatment. Nephrolithiasis complicated with RMC is mostly related to mechanical stimulation of kidney stones, chronic inflammation, and long-term stimulation of carcinogens in effusion. In such cases squamous cell carcinoma is more commonly observed relative to other carcinomas. The current described patient is an extremely rare case of severe obstructive hydrops with RMC caused by urinary tract stones; a clinical phenomenon that is rarely reported at home and abroad. Urinary tract calculi combined with RMC has an insidious onset and presents with atypical clinical symptoms that confound early diagnosis.

In this case, the main symptoms experienced by the patient were a pain in the left upper abdomen and back, with the presence of an obvious mass on the body surface. No definite renal space-occupying manifestations were found in the whole abdominal CT examination. Combined with the past medical history, the patient had undergone percutaneous nephrolithotripsy for left renal calculi with hydronephrosis, and it was considered that the renal structure might change due to the operation, likely the severe left renal hydrops with infection due to ureteral obstruction. To relieve the persistent pain, a laparoscopic left nephrectomy was performed with surgeons. Clinical studies have shown that although RMC is resistant to radiotherapy, chemotherapy, and immunotherapy(7), radical nephrectomy can still achieve better survival in patients with an early stage of disease and low metastatic burden. However, due to severe hydronephrosis, there is no specific basis for preoperative diagnosis, and tumor cells may be disseminated and implanted during the operation. In this case, the patient developed retroperitoneal metastasis and recurrence after surgery from which they did not recover and died shortly after.
Previous studies have shown that RMC is a rare tumor that is more commonly diagnosed in African American adolescents with sickle cell hemoglobinopathies. In contrast, in Chinese RMC patients, the occurrence of the sickle cell trait (SCT) is rare, and the occurrence of the disease is more common in middle-aged and elderly people. Only 9 cases have been reported in China, the youngest patients being 11 years and the oldest being 77 years (average: 42.8 years). In China, RMC affects more females than males (male:female = 0.8:1), and SCT was found in only two cases (9). The current case report describes a patient that was 64 years, which is significantly older than international RMC reports. In 2014, Amin (10) et al proposed the concept of "RMC unclassified renal cell carcinoma" for patients with an RMC immunohistochemical profile, but no SCT. This entity is much rarer than typical RMC and occurs in elderly patients and is based on a small number of case reports. In these cases, unclassified RCC with medullary features” or “RCC unclassified, with medullary phenotype” (RCCU-MP) are described to mostly occur in non-Africa ethnicities.

Preoperative diagnosis of urinary tract stones with renal malignancies, especially in the context of rare types of tumors, is difficult. First, both urinary tract stones and kidney tumors may have symptoms such as hematuria and lower back pain, which lack specificity. Thus, the symptoms of kidney stones can mask the symptoms of secondary or accompanying malignant tumors. Second, the presence of urinary tract stones combined with obstruction and renal tumors can cause changes in renal structure. In this case, the CT images of the patient were atypical. Additionally, the patient underwent ipsilateral percutaneous nephrolithotomy, which increased the difficulty of imaging diagnosis. Third, urinary exfoliated cytology is an important diagnostic method for renal malignant tumors, however, urinary tract obstruction and the accumulation of water make it difficult for tumor cells to be excreted with urine, reducing the success rate of urinary exfoliated cytology.

RMC is a rare and highly aggressive malignancy that even without a history of SCD or SCT, has rarely been reported in people of Asian descent. There is no specificity in imaging and the diagnosis is mainly based on histopathological parameters. Furthermore, treatment methods are limited. In our case, the preoperative diagnosis was unclear, and the treatment was delayed due to the combination of urinary tract stones and severe urinary tract obstruction. For elderly patients with long-term renal stones, hydrops, and altered kidney morphology, physicians should be extremely vigilant about the possibility of renal tumors. Preoperative CT or MRI enhanced examination may help reduce the rate of missed diagnosis and improve patient outcomes. When necessary, a biopsy may be required, however, this should be based on the patient's condition. Early detection, diagnosis, and comprehensive treatment are essential for improving the prognosis of RMC patients.

**Abbreviations**

RMC: Renal medullary carcinoma; SCD: sickle cell disease; SCT: sickle cell trait.

**Declarations**
Consent for publication

Written informed consent was obtained from the patient. A copy of the consent form is available for review and can be provided on request.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

n/a

Competing interests

The authors declare that they have no competing interests.

Funding

No funding was received for this study.

Authors' contributions

YQW wrote the manuscript and conducted the literature review. WYF, YLYLX and WJ participated in the clinical care of the patient. ZXM assisted in interpreting the results under clinical prospective. All authors assisted the results interpretation and manuscript revision. All authors read and approved the final manuscript.

Acknowledgements

Not applicable.

References


Figures
Figure 1

CT scan of the whole abdomen.
Figure 2

Immunohistochemistry of the resected left kidney showed that cancer cells was positive for AE1/AE3, Vimentin, CD10 and partially positive for CD117 and weakly positive for TFE3, Mitf. The INI-1 was negative and the Ki-67 index was 70%.