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Case Report / Olgu Sunumu

An Extreme Rare Case of Hematuria: Kidney Rhabdomyosarcoma in Elderly Woman

Son Derece Nadir Bir Hematüri Olgusu: Yaşlı Kadında Böbrek Rabdomiyosarkomu

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ABSTRACT

Hematuria is a warning and serious symptom of the disease with a broad spectrum. Although it first brings to mind diseases related to benign pathologies at young ages, it is one of the clinical markers for cancer, especially in the elderly and requires further investigation. There are some risk factors for cancer-induced hematuria such as male gender, age >35, analgesic abuse, chronic hemodialysis, radiation exposure or carcinogens. Additionally, rare diseases should not be ignored in the differential diagnosis of hematuria. Here, we present an extremely rare case of renal rhabdomyosarcoma of kidney in an elderly female patient who was admitted to the emergency department with hematuria.

Keywords: Hematuria, rhabdomyosarcoma, radical nephroureterectomy, metastasis to tonsilla

ÖZ

Hematüri, geniş bir spektruma sahip hastalığın uyarıcı ve ciddi bir belirtisidir. İlk akla genç yaşlarda iyi huylu patolojilere bağlı hastalıkları getirse de özellikle yaşlılarda kanserin klinik belirteçlerinden biridir ve daha fazla araştırma gerektirmektedir. Erkek cinsiyet, yaş >35, analjezik kötüye kullanımı, kronik hemodiyaliz, radyasyona veya kanserojenlere maruz kalma gibi kansere bağlı hematüri için bazı risk faktörleri vardır. Ayrıca hematürinin ayırıcı tanısında nadir görülen hastalıklar da göz ardı edilmemelidir. Burada, acil servise hematüri ile başvuran yaşlı bir kadın hastada böbrekte son derece nadir görülen renal rabdomiyosarkom olgusunu sunuyoruz.

Anahtar Kelimeler: Hematüri, rabdomiyosarkom, radikal nefroüreterektomi, tonsilla metastazı

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INTRODUCTION

Hematuria is the presence of blood in the urine.[1] It is divided into 2 types; microscopic that is invisible to the naked eye and gross which is seen.[2] It is mostly related to benign conditions such as infection, trauma, vigorous exercises. But some serious reasons also come into question as cancer of the bladder or kidney, infectious inflammatory diseases of the urinary tract organs, hematological diseases, coagulation disorders, and polycystic kidney disease. These serious diseases are more common in elderly patients with hematuria and are needed further clinical investigations for diagnosing.[3]

There are some risk factors for cancer-induced hematuria such as male gender, age >35, analgesic abuse, chronic hemodialysis, radiation exposure or carcinogens. However, in further clinical studies, hematuria due to very rare

causes can sometimes be detected. When hematuria is related to the upper urinary tract, it may be accompanied by a thread-shaped clot.

Furthermore, sarcomas constitute 1-3% of primary renal tumors.[4] Rhabdomyosarcoma (RMS) of the kidney is a subtype of renal sarcomas that is extremely rare. RMS of kidney arises from skeletal muscle progenitor cells. Although RMS is mostly seen in the pediatric population, they are uncommon in adults. Alveolar, embryonal, spindle cell, and pleomorphic are the subtypes of RMS.[5] However, approximately 25% of adolescent embryonal RMS (ERMS) occur in the genitourinary system, adult genitourinary RMS is scarce.[6] The small number of cases on this subject may also affect the effective use of treatment options after pathological diagnosis.

Here, we present an extremely rare case of adult ERMS as the cause of hematuria, in an elderly woman.

CASE REPORT

A 62-year-old female patient admitted to the emergency department of our institute with chief symptoms of gross hematuria and left flank pain. Neither she had comorbidities nor had a previous operation before. She had mild left flank pain in detailed physical examinations. The patient smoked for 40 years, quit 10 years ago. There was macroscopic hematuria in the urinalysis and mild anemia in the laboratory. There was a 10 cm diameter mass in the upper pole of the left kidney in ultrasound. There was no metastasis to the lung or other organs on computed tomography (CT) (Figure 1A, B).

The tumour was arising from the pelvis and was completely infiltrating the left kidney with necrotic areas. There was also a 4.5 cm diameter necrotic lymph node in the left paraaortic area.

After taking signed consent forms, we made cystoscopy and laparoscopic left radical nephroureterectomy. There was not any tumour in the bladder. We made plaque surgical technique and resected the left ureter during cystoscopy. Additionally, we performed laparoscopic retroperitoneal lymphadenectomy simultaneous. Because of the tumour was infiltrating left adrenal gland, it was also resected.

There was not any complication during operation and clinical follow-up. She was discharged 5th day after the surgery.

There were spindle cells in the fascicular arrangement with extensive necrosis in pathology examinations. Pathology reported ERMS of the kidney (Figure 2). The tumour was infiltrating the adrenal gland but the surgical margins were negative. Additionally, all lymph nodes were reactive without metastasis.

DISCUSSION

Hematuria has a broad spectrum. However, rare diseases should not be ignored, in the differential diagnosis of hematuria is not the subject of this case report. As a rare disease, primary renal sarcoma constitutes 1-3% of all kidney tumors.[7] Its diagnosis is based on the origin of the tumour from renal parenchyma, exclusion of metastatic sarcoma and the sarcomatoid subtype of kidney cancer.[7] The genitourinary RMS mostly affects the bladder or pelvic organs of children aged 2-6 years.[8] The RMS of the kidney is an extremely rare entity in adults. Fang et al.[9] reported the existence of 11 cases in adults in 2014. In 2014, Fanous et al.[5] published a case of primary renal ERMS in a 37-year-old woman.[9] According to our best knowledge the 14th case of ERMS of kidney in the literature.

ERMS accounts for almost 66% of all RMS cases and may have a better prognosis. Nevertheless, recommendations that can be used in the diagnosis and treatment of this disease cannot be made. However, ERMS is aggressive cancer and its clinical manifestations are similar other primary sarcomas.

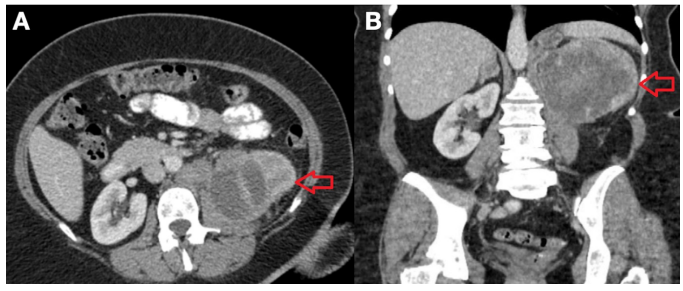

Figure 1. Enhanced computed tomography slides of the renal mass. A) Arrow show the left kidney mass in axial slide, B) Arrow show the mass in coronal slide

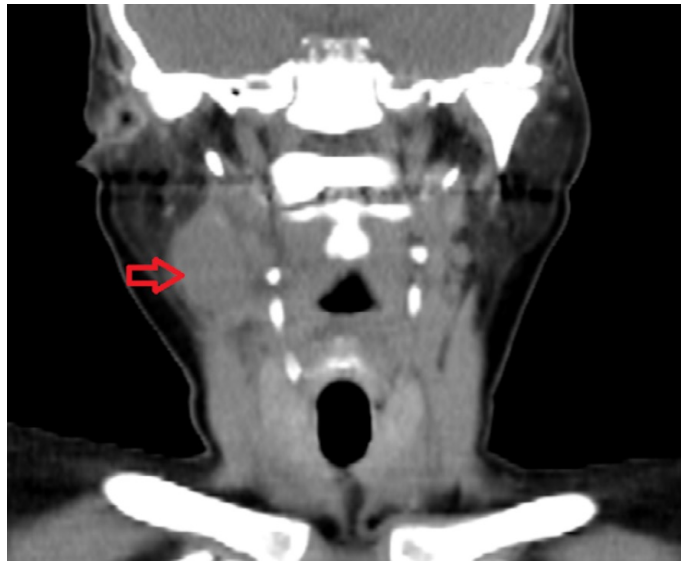

Figure 2. Coronal slide of neck. Tonsilla metastasis of embryonal rhabdomyosarcoma of left kidney

Large non-specific soft-tissue masses with poor contrast enhancement are the imaging characteristics of sarcomas that these are indistinguishable from renal cell carcinoma/upper urinary tract transitional cell cancer.[10] Therefore, it is difficult to talk about the distinguishing feature of ERMS in contrast-enhanced computed tomography examination. After all, the differential diagnosis of ERMS is difficult with clinical and imaging findings. The histopathological examination makes the definitive diagnosis. Although this can be a challenging needed experience. Myogenin and MyoD1, myogenic regulatory proteins expressed early in skeletal muscle differentiation, are considered sensitive and specific markers for RMS and are more specific than desmin and more sensitive than myoglobin.[11]

Because of the ERMS of kidney is an extremely rare entity, to draw a standard the follow-up is very difficult. We performed positron emission tomography after pathology report, 30th day of surgery. There was a mild activity in neck region tonsillar area. Additionally, the patient had sore throat. We referred the patient to the otorhinolaryngology department. After the physical examination, the patient undergone neck CT. there was a mass in the tonsillar region (Figure 2). The biopsy of the tonsil was reported metastasis of ERMS in pathology.

We know that primary renal sarcomas have a poor prognosis and metastasis occurs at diagnosis in 90% of cases.[12] The basis of treatment continues to be radical nephrectomy. Hawkins et al.[13] reported that adult RMS has a worse prognosis than in children as other publications on this issue. Furthermore, the relationship shown between the RMS subtype in children and survival has not been demonstrated in adults.[13]

However, there was not any metastasis, we diagnosed tonsillar metastasis of ERMS, in this case (Figure 3A, 3B). This is another rare entity as the first case of ERMS with tonsillar metastasis as the first case in the literature. The patient is getting chemotherapy at the oncology department of our institute nowadays.

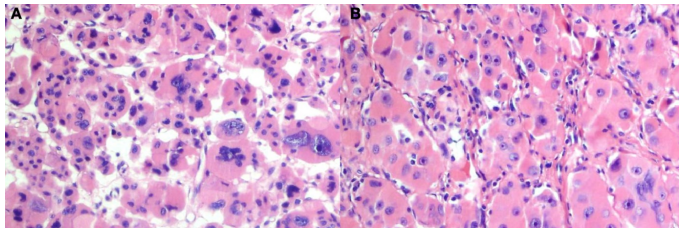

Figure 3. Pathological slides of the tumour. A) Widespread necrosis areas (H&E, x10), B) Fields of spindle cells in the fascicular arrangement (H&E, x20)

CONCLUSION

The primary ERMS of the kidney is extremely rare in adults. This aggressive tumour can metastasize in a short time and cause a poor prognosis. Thus, hematuria should be well researched notably in elderly patients. Unfortunately, there has not been a well-established chemotherapy regimen for ERMS of the kidney.

Ethics

Informed Consent: Signed informed consent was taken from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Y.A., O.K., K.D., A.F.B., Concept: Y.A., S.N.G., F.E.T., Design: O.K., F.H.D., E.M.Y., Data Collection or Processing: K.D., M.K., Analysis or Interpretation: A.F.B., M.K., F.E.T., Literature Search: S.N.G., F.H.D., E.M.Y., Writing: Y.A., K.D.

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