



Böbreğin oldukça nadir görülen çocukluk çağı tümörü, Renel Clear Cell Sarkom: Olgu Sunumu

Extremely Rare Childhood Tumor, Renal Clear Cell Sarcoma: A Case Report

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ABSTRACT

Clear-cell sarcoma of the kidney (CCSK) is a rare tumor and comprises 3% of primary pediatric malignant renal tumors. It is known as an aggressive tumor that metastases to the bones with a poor prognosis. Clinically and radiologically, it can mimic Wilms' tumor. We present a CCSK case in the left abdominal area in a 12 year old patient.

Key Words: Clear cell sarcoma; Wilms tumor; Child

ÖZET

Böbreğin Clear-cell Sarkomu çocukların primer malign tümörlerinin yaklaşık %3'ünün oluşturur ve nadir görülen bir tümördür. Kemiklere metastaz yapması ve kötü prognoza sahip olması nedeni ile saldırgan bir tümör olarak bilinir. Klinik ve radyolojik olarak Wilms' Tümörüne benzer. Karın sol boşluğunda yerleşen 12 yaşında olguyu sunduk.

Anahtar kelimeler: Clear Cell Sarkom, Wilms Tümörü, Çocuk.

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INTRODUCTION

Clear-cell sarcoma of the kidney (CCSK) is a highly malignant neoplasm occurring in the same age range as Wilms' tumor. CCSK is a rare malignant tumor of childhood, known for its aggressiveness, its tendency to recurrence and to metastasis to bone (1). CCSK is an uncommon renal malignancy of childhood, representing about 3% of pediatric renal tumors (2, 3). Age of presentation ranges from 2 months to 14 years, with a mean age of 36 months (4).

We report a pediatric case aged 12 years carrying a large abdominal mass. The aim of the present report is to present a rare CCSK age with distant metastasis but with local recurrence. As it shows radiological and histopathological similarities with other renal tumors care is heavily emphasized.

Case Report

A 12 year male patient had applied two years ago to the pediatric clinic with severe upper left quadrant abdominal pain and due to the presence of large amount of blood in his urine. During the radiological examination a 110x90cm mass was detected on the left kidney. The patient was referred for further examinations to the Pediatric Oncology Clinic and the following were diagnosed; WBC:9,800 HGB:10,6 PLT:247,000 CRP:10,1 Procalcitonin: 0,09. The abdominal tomography made revealed left renal originating 9x10 cm mass, left renal vein thrombus, and

1x1 cm left paraaortic lymphadenopathy. According to the radiological findings, Wilms' tumor possibility had been reported (Figure 1). Hence, in line with the preoperative treatment protocol of Turkish Pediatric Oncology Group (TPOG) in Wilms'tumor vincristine 1.4 mg/m² and 0,015 mg/kg Actinomycin D was administered. As the mass was large, surgery was planned after chemotherapy to shrink the mass. Since patients response to chemotherapy was inadequate and no shrinkage in patients mass, patient underwent surgery right after chemotherapy. Since the pathology report was CCSK (Figure 2, 3) the oncologic chemotherapy protocol was administered interchangeably (doxorubicin, cyclophosphamide, and cyclophosphamide and vincristine, etoposide) 1



Figure 1. 9x10 cm mass with left renal origin tomography.

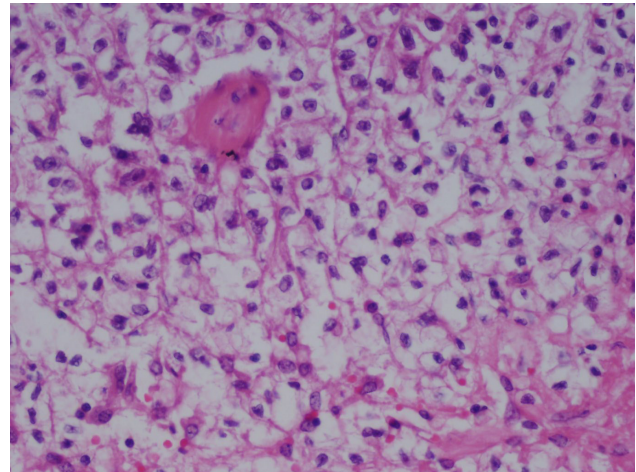


Figure 2. Clear cell karsinom H&E x400 Magnifying.

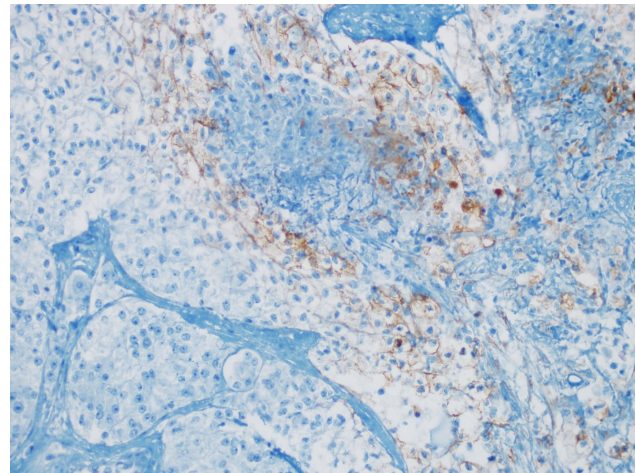


Figure 3. Immunohistochemical staining for Epithelialmembraneantigen (EMA).

month after chemotherapy was applied. After a 1 year therapy, in the 4th month of the routine controls a Pet-CT scan was made due to the presence of metastatic lung nodules in thoracic computerized tomography. Nodular lesions without fluorodeoxyglycos A (FDG) uptake were reported for both lungs. The MR images showed increased FDG uptake in the left kidney indicating recurrence. The patient with good overall assessment findings, was re-operated after 18 months in order to prevent recurrence driven obstructive problems and relapse tissues were removed and signs using titanium clips were placed. During the post-operative follow up, lesions progression in the lungs was seen in the tomography images and chemotherapy reinitiated. Patient is currently treated in the oncology service.

DISCUSSION

Clear cell sarcoma also known as “Bone Metastasizing Tumor of Childhood” comprise 3% of all primary childhood renal tumors (5). Approximately 5% of patients have metastatic disease at presentation. However in the present case there was no bone metastasis. The most common site of metastasis at the time of presentation in patients with CCSK is the ipsilateral renal hilar lymph nodes. Treatment consists of chemotherapy and nephrectomy with current long-term survival rate of 60-70% (6). In these cases, in order to determine the stage

during the treatment lymph node specimens are required.

Sonography is the initial modality to evaluate abdominal mass and to demonstrate renal origin of the tumor. Imaging studies are unfortunately entirely nonspecific (7). Clinical and imaging modality characteristics may support a particular diagnosis. The pretreatment gold standard is histopathologic evaluation.

CCSK is a malignant mesenchymal neoplasm that includes undifferentiated cells, cords and nests separated by fibrovascular septa, and abundant extracellular matrix. Nevertheless, there are no tumor specific markers for CCSK, which makes the diagnosis difficult. Clear cell sarcoma of the kidney is a rare aggressive tumor with nonspecific imaging findings. The radiologist should be aware of CCSK and include it in the differential diagnosis of pediatric renal tumors.

Differential diagnosis is an essential clinical entity in CCSK as it can be confused with other malignant kidney tumors, show extremely poor clinical outcome, and has a high metastasis and relapse risk. Although imaging methods give accurate results, these have to be verified with histopathological examination. As in the present case, CCSK confused radiological with Wilms' tumor in children might be the case. As it will change the diagnosis and hence pre and post-surgery treatment protocols, its' reflection in the prognosis will have a positive impact.

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