

# Clear Cell Sarcoma of Kidney: A Rare Pediatric Renal Tumor

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## ABSTRACT

Clear cell sarcoma of the kidney is usually diagnosed on sixth or seventh month after the birth and the second peak is on two or three years old. In this report, clear cell sarcoma of the kidney detected at a 13-years-old boy is presented with its histopathologic aspects.

**Key words:** Sarcoma, clear cell, kidney

## Böbreğin Berrak Hücreli Sarkomu: Nadir Bir Çocuk Renal Tümör

### ÖZET

Böbreğin berrak hücreli sarkomu genellikle doğumdan sonra altıncı veya yedinci ayda teşhis edilir ve ikinci zirve, iki ya da üç yaşındadır. Bu raporda, 13 yaşındaki bir çocukta tespit edilen böbreğin berrak hücreli sarkomunun histopatolojik yönleri sunulmaktadır.

**Anahtar kelimeler:** Sarkom, berrak hücreli, böbrek

## INTRODUCTION

Clear cell sarcoma of the kidney (CCSK) also called a "bone-metastasizing renal tumor of childhood" constitutes 3% of renal tumors in childhood and is the second common pediatric renal neoplasm (1). CCSK is usually diagnosed on sixth or seventh month after the birth and the second peak is on two or three years old. CCSK of adoloscence or adults is extremely rare (2). In this report; CCSK detected at a 13-years-old boy is presented with its histopathologic aspects.

## CASE

A 13-year-old boy presented with a 2-month history of left flank pain. Abdominal and pelvic computerized tomography showed a 6x5 cms size heterogeneous mass originating from the lower pole of the left kidney. A left radical nephrectomy with hilar lymphadenectomy was performed. On macroscopic examination; the mass was measured to be 6x5,5x4 cms, had irregular contours

and gray-yellow, gelatinous cut surface. Microscopic evaluation revealed the tumor composed of nests and cords and separated from kidney paranchyma with fibrovascular septas. Tumoral cells had oval, round nuclei with granular chromatin and eosinophilic or clear cytoplasm with indistinct borders (figure 1). There was some atypical mitoses within the tumor. The histopathological diagnosis was clear cell sarcoma of the kidney. No lymph node metastases were found. Concomitant chemo-radiotherapy was performed. Further evaluation including bone scan did not demonstrate any evidence of metastases. There was no evidence of local recurrence or metastases during the following twenty months after therapy.

## DISCUSSION

Clear-cell sarcoma of the kidney is a rare pediatric renal sarcoma with poor prognosis and propensity to metastasize to bone. It is a distinctive renal malignancy regarded as a morphologic feature of Wilms' tumor (3). It has

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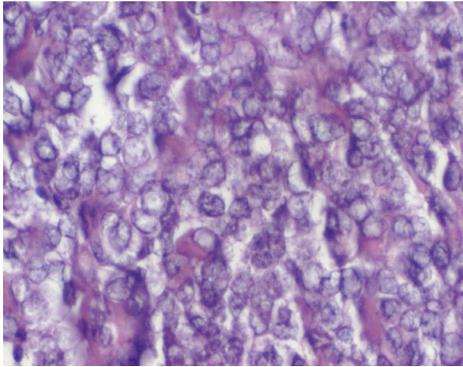
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**Figure 1.** Tumoral cells has oval, round nuclei with granular chromatin and eosinophilic or clear cytoplasm with indistinct borders (HEX400).

an aggressive evolution with a high rate of recurrence and mortality (2) and a wider distribution of metastases than Wilms' tumor (1). Zigman et al. (4) reported a case of CCSK with transvenous tumor extension into the right atrium. Hung (5) reported a hydropic male fetus died in utero with metastatic disease from a renal clear cell sarcoma at 31 weeks gestation. The tumor had metastasized to para-aortic and mediastinal lymph nodes, lung, pleura, and liver, leading to superior vena cava obstruction and pulmonary hypoplasia. In the literature; CCSK is reported as a disease of infancy or early years of life (2). The detection of CCSK in a 13-years-old patient is the distinctive feature of our case.

El Kabari et al. (1) suggested that early diagnosis of CCSK is needed for an adapted treatment with regards to its aggressiveness and its ability to give bone metastases. In our case; no metastasis was detected both on the first presentation and on the follow-up period of twenty months.

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