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Clear cell sarcoma of the kidney in a newborn

Bir yenidoğanda böbreğin clear cell sarkoması

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Abstract

In the present article, we present the case of a 37-month-old male newborn, who was delivered by normal birth with no history of routine examinations during pregnancy, with an abdominal mass observed after the birth, and was eventually diagnosed with clear cell sarcoma of the kidney in the histopathology examination after the mass was dissected by surgery. We report the observed symptoms as well as related cases found in the literature related to childhood and newborn age clear cell sarcoma of the kidney. We also put forward clinical observance, stages, management and outcomes of clear cell sarcoma of the kidney during newborn. Our study underlines that clear cell sarcoma should be kept in mind during differential diagnosis although it is rarely observed in newborns.

Keywords: Clear cell sarcoma; Kidney; Newborn; Children.

Öz

Bu makalede, 37 haftalık, gebelik süresince herhangi bir sağlık kurumunda tıbbi takibi olmadan normal vajinal yolla doğan, doğumun ardından yenidoğan ünitesinde karnında kitle fark edilen bir erkek yenidoğan olgu sunuldu. Cerrahi olarak çıkarılan kitlenin histo-patolojik incelemesinde kitlenin böbrek kaynaklı clear cell sarkoması olduğu tesbit edildi. Olgudaki mevcut bulgular sunulup, böbreğin clear cell sarkomasının çocukluk çağı ve yenidoğan döneminde ortaya çıkışı ile ilgili literatür araştırması yapılarak bununla ilgili vakalar bildirildi. Yenidoğan döneminde böbreğin clear cell sarkomasının klinik olarak ortaya çıkış şekilleri, evresi, yönetimi ve sonuçları anlatıldı. Böbreğin clear cell sarkomasının yenidoğan döneminde oldukça nadir görülmesine rağmen ayrıcı tanıda akılda tutulması gerektiği vurgulandı.

Anahtar Kelimeler: Clear cell sarcoma; Böbrek; Yenidoğan; Çocukluk.

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INTRODUCTION

Renal tumors are rarely seen in newborns and fetuses (1-2). One of these tumors, clear cell sarcoma of the kidney (CCSK), which is also known as renal sarcoma with bone metastasis, is among aggressive neoplasms. It is usually diagnosed around age 3 and its prevalence is twice as much in males. It is very rarely seen during the first 6 months of life and it is again very rarely diagnosed congenitally. CCSK are metastases to bones, liver, lungs, and lymph nodes. Extra-renal clear cell sarcoma of the kidney has the same morphology and phenotype (3).

This is a presentation of a case of a male subject with an intra-abdominal mass observed immediately after birth without no determined metastasis after the examinations who was operated on post-natal 5th day. The mass was dissected and as a result of the histopathology examination of the mass, it was diagnosed as renal clear cell sarcoma. In the literature review conducted, it was found that 7 other cases, two of which were extra-renal cases, were investigated previously.

CASE REPORT

Our male patient was born as the first child of a 21-year-old mother in the 37th gestational week by normal vaginal delivery with a weight of 3100 g in the hospital. There were no complications during the birth and the subject was assessed normal after birth. Mother did not receive any routine medical examination during her pregnancy. An abdominal mass was observed in the subject after the birth. Physical examination displayed stable vital signs. On abdominal examination, a hard, immobile mass that approached the midline from hard flick lateral was palpated at the left upper quadrant (Figure 1).

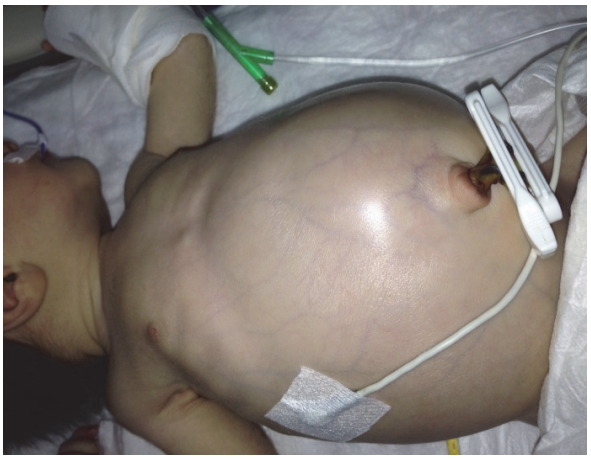

Figure 1. Preoperative outlook of the patient with distension in the abdominal upper left quadrant.

Hematological laboratory biochemical test results were normal except for wbc: 15.3 K/mm^3, hgb:10.7g/dl and indirect bilirubin: 2.6 mg/dl. In the radiological examination of the abdominal cavity (USG), a vascularized 8 x 9 x 10 cm mass lesion of the left kidney that filled the left half of abdominal cavity, including cystic and necrotic areas was observed. In mass posterior, parenchymal tissue of the left kidney was identified. Abdominal computed tomography showed a lesion of 9 × 9.2 × 8.8 cm with heterogeneous density that partially crossed the midline, displacing the midline vascular structures to the right side (Figure 2). A metastasis of the brain, thorax or bones, or other intra-abdominal organs was not identified.

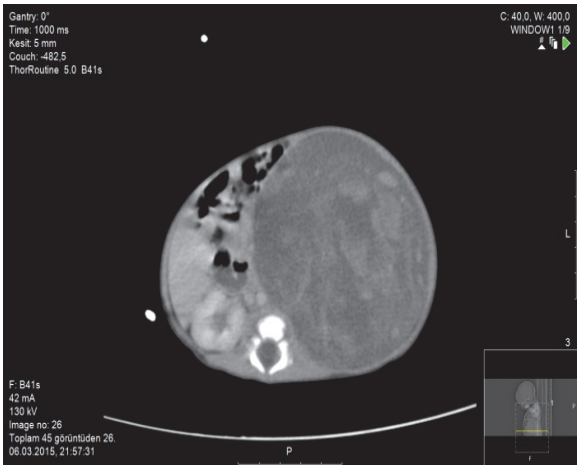

Figure 2. Computed tomography revealed a mass of heterogeneous density that partially crossed the midline, pushing the midline vascular structures to the right side.

The patient was operated on post-natal 5th day. A consultation was conducted before the surgery and chemotherapy was ruled out since the type of the tumor was unknown. During the surgery, it was observed that the dimensions of the left renal mass was 9 x 8 x 7 cm and it was well circumscribed and partially mobile with an intact capsule and did not invade beyond the midline, pushing vascular structures towards the medial. It had a normal outlook and was not attached to the left ureter periphery or was not related to the mass. Left kidney parenchyma was identified at the lower pole of the mass and was uncircumscribed. Left adrenal gland was at the upper pole of the mass and was uncircumscribed. The capsule of the mass was attached to the retroperitoneal area at renal vascular structures but was incised by separating it from the surrounding tissues and without leaving behind a residue. No affected lymph nodes were identified in the area.

Macroscopically, the mass weighed 300 g, 10 × 7.5 × 6 cm in size, and was encapsulated. Before the removal, the mass surrounded the ureter, the left kidney, and the left surrenal gland. Approximately 70% of the mass consisted of necrotic areas.

Histopathological examination revealed diffuse neoplastic proliferation of cells with oval to spindle-shaped nuclei (some of the nuclei were vesicular) and clear to slightly eosinophilic cytoplasm. A small number of bizarre-shaped nuclei were also observed (Figure 3a). Twelve mitoses were observed in 10 high-magnification areas (Figure 3b). The tumor was identified as a 'typical clear cell sarcoma' (Figures 3b and 3c).

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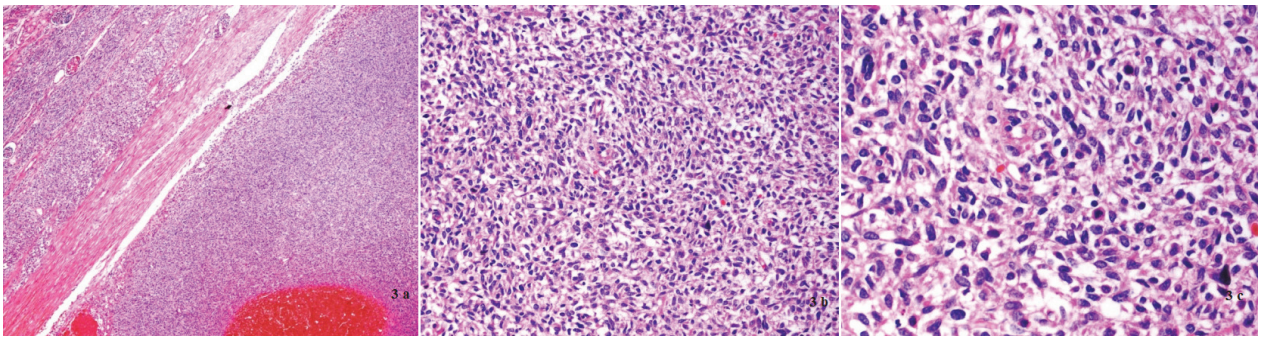

Figure 3a. Diffuse neoplastic proliferation consisting bizarre neoplastic cells (Hematoxilen-Eozin, x 200), **Figure 3b.** Mitotic figures (arrows) (Hematoxilen-Eozin, x400), **Figure 3c.** Neoplastic proliferation crossover the pseudocapsule that envelops the tumour with undamaged glomeruli within (Hematoxilen-Eozin, x100).

Immunohistochemically, vimentin (Dako, clone V9) diluted to 1:500, CD99 and EMA clones (E29, 1:100), pan-cytokeratin (MNF 116, Dako, 1:100), and desmin (Dako, clone D33, 1:100) antibodies were introduced to the tumor tissue. The immunohistochemical examination results demonstrated the following findings: desmin, negative (Figure 4a); SMA, negative; pan-cytokeratin, negative; CD99, diffuse 2+ (Figure 4b); WT, nonspecific (Figure 4c); and vimentin, diffuse 3+ (Figure 4d).

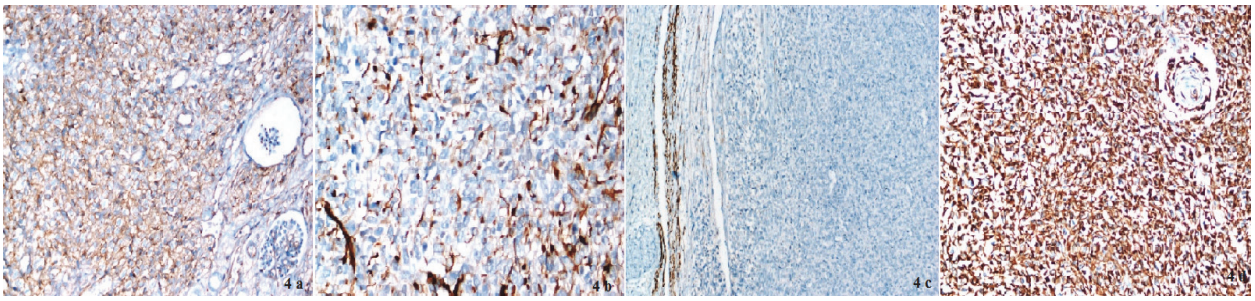

Figure 4a. Desmin negative, x 200, **Figure 4b.** CD-99 positive, x 200, **Figure 4c.** WT1 negative, x 400, **Figure 4d.** Vimentin, diffuse strong positive, x 200.

With the existing findings, it was determined that there was no distant organ metastasis, no regional lymph node involvement; the mass extruded the kidney but was removed totally with the kidney, and was Grade-2 since there was no tumor tissue left within the limits of nephrectomy.

Postoperative chemotherapy was not administered. The patient was discharged on postoperative 7th day. Treatment of the patient was conducted at another health center on the request of the patient's family. Currently, the patient has no health problems and is 11-month-old. Follow-up examinations demonstrated no sign of relapse.

DISCUSSION

Pediatric renal tumors are the second most prevalent tumors among abdominal malignancies in infants and children. Eighty-five percent of these are Wilms' tumor, 5% are congenital mesoblastic nephroma, 5% are CCSK and 2% are rhabdoid tumor of the kidney (4). Peak incidence of the CCSK is between 2-4 ages and male to female ratio is 2:1. Histologically classical pattern is the most prevalent (90%) and among the rest 50% are myxoid, 35% sclerosing, 26% epitheloid, 11% palisading, 7% spindle-cell, 4% storiform, and 3% are anaplastic (5).

Mean incidence age is 36 months. They are generally detected by abdominal distention or abdominal mass clinical complaints. They could be indicated by abdominal pain and gross hematuria (5). Literature review would demonstrate that there were only a few cases below the age of 6 months and there was only 1 case identified in the intrauterine period (3 – 10, 11).

Characteristic clinic symptoms of newborn CCSK cases are abdominal distention and detection of intra-abdominal mass with palpation. Furthermore, it could present with jaundice, vomiting, and stillbirth rarely (12). In addition, it could be detected by abdominal USG (3).

In studies conducted on CCSK, chromosome translocation and other genetic variations were identified. Molecular biological pathogenesis of these variations is still unknown. Its action on prognosis is also unknown. The most frequently observed change was loss of 19p (8 cases) and the gain of 1q (5 cases) (5). In a genetic research conducted with cases younger than 6 months, Hung detected a translocation between chromosomes 13 and 14 (3). Punnet et al. found a translocation between 10 and 17 (13).

Immunohistochemically, CCSK displays consistent nonspecific positivity for vimentin. Today, nerve growth

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factor receptors (NGFR) were also identified immunohistochemically. Cytokeratin, Mic-2, S100, neural markers and desmine and WT-1 are uniformly negative in CCSK (5). Satoh et al., in their case of an infant younger than 6 months, noted CD10 positivity in the related kidney and clear cell sarcoma of the kidney (14). In the Hung case, CD10 was determined in the tumor as strong positive focally and diffused vimentine was immunohistochemically determined as strong positive (3). In the present case, immunohistochemical examination demonstrated CD99: Diffused 2+ and Vimentine: Diffused +3 positive.

Literature review demonstrated that there were 7 more cases younger than 6 months who were diagnosed with renal clear cell sarcoma and 2 of these were located in extra-renal locations (Table 1) (3, 6 – 8, 11, 15).

Mean gestation ages of these cases were 39 weeks (31 weeks – 43 weeks). Three were diagnosed in the 1st stage, 2 were in the 2nd stage, and 2 were in the 4th stage. The mean weight of these tumors was 180 g (15-320 g). The mean birth weight of the babies was between 2340 g and 3800 g (12). Renal clear cell sarcoma was detected in the intrauterine in only 1 case (3). National Wilms Studies report 2 cases that were younger than 6 months (4). Four additional cases were found separately in the literature and 3 of these were newborns and only 1 had intrauterine tumor (3, 6, 9, 10). Most of these cases were males. Interestingly, both extra renal localized cases were females (10, 15). In none of these cases were there cystic areas around the tumor and, the mass contained wide necrotic and cystic regions. In two cases with metastasis, the metastases were to para-aortic and mediastinal lymph nodes, lung, pleura and liver. Furthermore, superior vena cava obstruction was formed due to metastasis (3). CCSK originates in primitive cells in the kidney.

Extra renal localized ones originate from mesonephric residues (6). In our case, it originated in primitive kidney cells. Usually there are no macroscopic cystic areas around the tumor however they existed in our case.

Table 1. Clear cell sarcoma of the kidney as reported in the literature (A&W alive and well, DwD death with disease, PR peri renal, NC non-clasical, NS not specified, m months, NB newborn)

Author	Age	Sex	Site	Variant	Metastases	Follow up	Outcome
Present case	NB	male	Kidney	Classical	Negative/No PR invasion	11 m	A&W
Suzuki et al. 1983	NB	male	Kidney	Classical	Negative/No PR invasion	12 m	A&W
Mazzoleni et al.2003	NB	male	Kidney	NC	Negative/No PR invasion	25 m	A&W
Kataoka et al.1993	NB	female	Ileal valve	NS	Multiple/local	1 m	D&D
Newbould and kelsey.1993	4m	male	Kidney	Classical	PR invasion	NS	NS
Argani et al.2000	5m	female	Ovary	Classical	NS	NS	NS
El kabari et al.2004	5m	male	NS	NS	NS	NS	NS
Hung 2005	fetal	male	Kidney	Classical	Multiple	-	D&D

There are two late-detected newborn cases with multiple local invasion and both are lost due to the disease. The National Wilms Study (NWTS) determined the low stage group as favorable prognosis group. Three different treatment regimes are proposed by NWTS. These are: standard resection, standard resection + chemotherapy and standard resection + chemotherapy + radiotherapy (12). Preoperative chemotherapy is not implemented in newborns with CCSK diagnosis. The medicines used in chemotherapy are actinomycin D, vincristine, etoposide, cyclophosphamide, and doxorubicin. Non-relapse survival for 6 years was reported between 25% and 63% in CCSK patients (5).

In patients younger than 6 months, the prognosis of the disease was as follows: Kataoka et al. determined the tumor in a patient in stage 4, reported that exitus was rapid despite aggressive therapy and surgery and chemotherapy (8). Hung examined a 31-week intra-uterine exitus fetus and reported that the tumor originated in the upper pole of the right kidney (3). Our patient, Suzuki's patient and Mazzoleni's patient are still alive, but the prognosis of the remaining 3 cases is unknown. Suzuki et al. do not recommend and implement aggressive chemotherapy when this tumor is detected in premature patients, since favorable prognosis of renal neoplasia is dependent on sarcomatous type (11).

These cases demonstrate that clear cell sarcoma of the kidney should always be considered in differential diagnosis though it is rare among intra-renal and extra-renal congenital tumors. Appropriate radiological examination should be conducted to assess especially brain and bone metastases and preoperative blood preparation while necessary liquid electrolyte support should be provided. Also, the incision should be wide enough for the resection of the mass in the treatment, sufficient liquid support should be provided during the surgery, and the tumor should be resected totally with radical nephrectomy and should be followed by postoperative chemotherapy and radiotherapy. Today, significant improvements are identified in the prognosis of CCSK tumors with multi-agent chemotherapy regimes and radiotherapy programs (5, 14).

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