

CASE REPORT / OLGU SUNUMU

## Surgical management of clear cell sarcoma of the kidney with tumor thrombus in the inferior vena cava and lung metastasis

### *Pulmoner metastaz yapmış ve inferior vena kavada tümör trombüsü olan renal clear hücreli sarkomun cerrahi yönetimi*

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#### ABSTRACT

Clear cell sarcoma of the kidney (CCSK) is a rare renal neoplasm of childhood. While CCSK is more common in children, it is extremely rare in adults. In CCSK, a tumor thrombus is sometimes formed in the inferior vena cava. Herein, we describe a rare case of an 18-year-old female with CCSK with vascular tumor thrombus extending to the inferior vena cava that was managed successfully by excision of the renal mass and vena caval thrombus, and pulmonary metastasectomy using cardiopulmonary bypass without cardiac arrest.

**Key words:** Renal Clear cell sarcoma, inferior vena cava, tumor thrombus, cardiopulmonary bypass

#### INTRODUCTION

Clear cell sarcoma of the kidney (CCSK) is a rare renal neoplasm of childhood. It is more common in children than in adults and is known as an aggressive tumor with poor prognosis. CCSK has a propensity to metastasize to lymph nodes, bone, lung and liver. Like Wilms tumor (WT), CCSK can also invade the inferior vena cava with extension into the right atrium. However, this is considerably rare. To our knowledge, only 3 adult and 6 pediatric cases of CCSK with inferior vena caval or cavo-atrial involvement have been reported in the literature to date [1,2].

#### ÖZET

Böbreğin clear hücreli sarkomu (BCHS) çocukluk çağıının nadir bir renal tümörüdür. Çocuklarda daha yaygın görülürken, yetişkinlerde oldukça nadir görülür. BCHS'da tümör trombüsü bazen inferior vena kavaya ilerleyebilir. Burada biz inferior vena kavaya yayılan vasküler tumor trombüslü 18 yaşındaki BCHS'lu nadir vakada kardiyopulmoner bypass altında ve kardiyak arrest yapılmaksızın renal kitlenin, vasküler tümör trombüsünün ve pulmoner metastazın başarılı olarak çıkarılmasını tanımladık.

**Anahtar kelimeler:** Böbreğin clear hücreli sarkomu, inferior vena kava, tümör trombüs, kardiyopulmoner bypass

In this report, we describe a rare case of an 18-year-old female with CCSK with vascular thrombus extending to the inferior vena cava that was managed successfully by excision of the renal mass and vena caval thrombus, and pulmonary metastasectomy using cardiopulmonary bypass (CPB) without cardiac arrest.

#### CASE REPORT

An 18-year-old female was admitted to our hospital with right sided abdominal mass. On physical examination, a large abdominal mass was palpated in the right side of the abdomen. Apart from slightly

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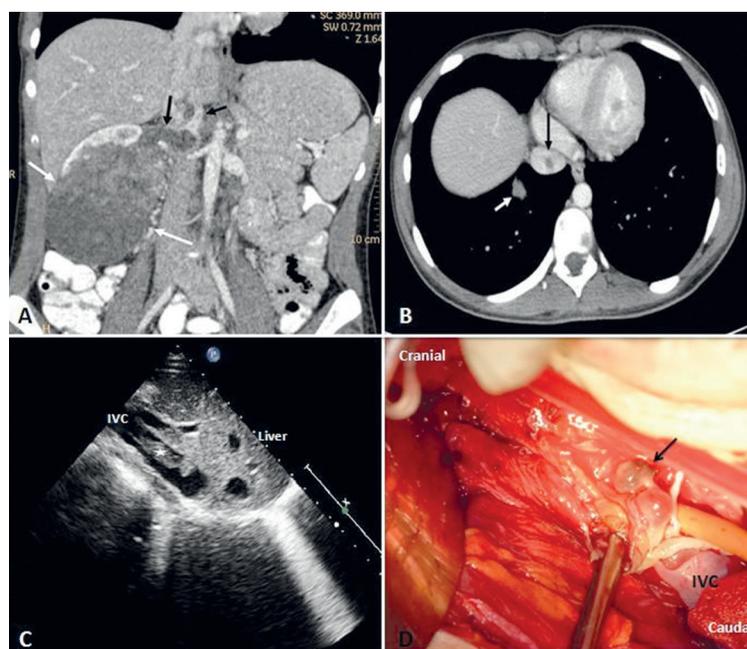
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elevated erythrocyte sedimentation rate of 25 mm/h (normal range 0-19 mm/h), no abnormality was noted in the other laboratory tests.

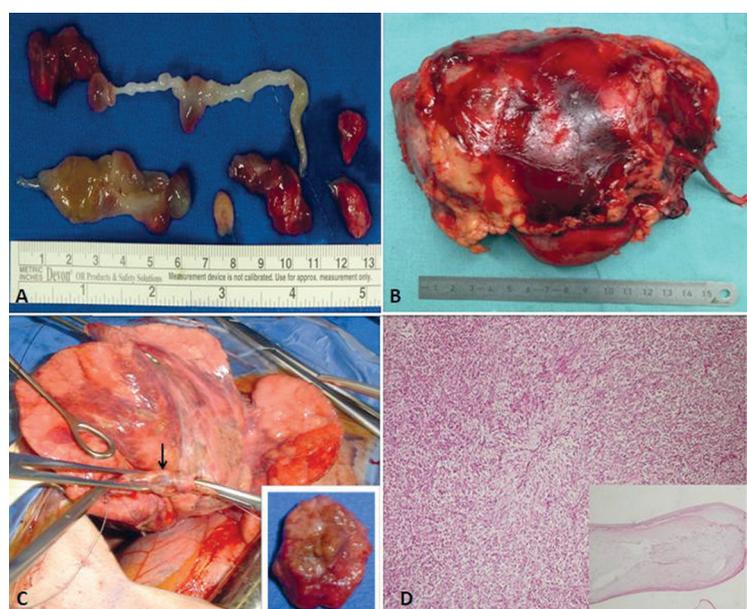
The contrast-enhanced computed tomography (CECT) of the abdomen and chest demonstrated a 15 x 12 x 8 cm (CC x ML x AP) heterogeneous solid tumor in the right kidney with vascular tumor thrombus extending through the right renal vein into the inferior vena cava (IVC) that the apex of the tumor thrombus (level III) existed just below the

inferior cavoatrial junction (Figure 1A) [3]. A single pulmonary metastasis in the right lower lobe was also noted (Figure 1B). Bone scintigraphy and brain magnetic resonance imaging revealed no further distant metastases.

Transthoracic echocardiography (TTE) showed tumor thrombus causing partial obstruction just below the cavoatrial junction in intrahepatic portion of IVC. The tumor thrombus extension into the right atrium was not detected (Figure 1C).



**Figure 1.** A) Heterogeneously enhancing solid mass occupying the renal pelvis and the lower pole of the kidney (white arrows) is demonstrated on coronal portal phase computed tomography image. Tumor thrombus filling the renal vein and extending to the inferior vena cava (IVC) is also depicted (black arrows). B) Tumor thrombus in the IVC (black arrow) is demonstrated on the axial portal phase computed tomography image through atrio-caval junction. Solid nodular mass on the right lung lower lobe anterobasal segment consisted with metastasis (white arrow) is also visible on this image. C) Echocardiography in subxiphoid view revealed the serpentine mobile tumor thrombus in the IVC just below the cavoatrial junction (The asterisk indicates tumor thrombus). D) Intraoperative photograph shows the removal of tumor thrombus (black arrow) in the IVC.



**Figure 2.** A, B, C) Intraoperative photographs show complete removed tumor thrombus, nephrectomy, and pulmonary metastasectomy specimen (The black arrow indicates pulmonary metastasis). D) Microphotograph shows that CCSK consisted of cells with fine nuclear chromatin, pale cytoplasm and indistinct cell borders forming nests separated by a fibrovascular stroma and mitotic figures are rare (HE, x20). Tumor tissue shows abundant myxoid stroma in the area of the renal vein invasion (small window, HE, x2).

After induction of general anesthesia, trans-esophageal echocardiography (TEE) was performed to confirm the proximal extent of the tumor thrombus. TEE was continued during the operation to detect tumor embolisms and to check complete extraction of the thrombus. It confirmed that the tumor thrombus extended to just below the cavo-atrial junction but not into the right atrium.

The surgical approach was obtained through a median sternotomy and laparotomy incision. Before heparinization, the laparotomy was performed and the left renal vein and infrarenal part of the IVC were encircled with the tape. Suprarenal part of the IVC was not encircled due to the risk of tumor thrombus embolization. Following sternotomy and heparinization, CPB was established through cannulation of the ascending aorta, superior vena cava, and right common femoral vein. CPB was initiated without cross clamping neither cardioplegic cardiac arrest.

Via a sternotomy, the intrapericardial IVC confirmed by TEE was found to be free of tumor and IVC at the cavoatrial junction was encircled with the tape. The intrapericardial IVC, infrarenal IVC and contralateral renal vein were then occluded with a tourniquet. The venotomy was started from the right renal vein and extended to the IVC. The tumor thrombus was partly adherent to the reno-caval junction, but it was not adherent and free floating in the IVC. The tumor thrombus was completely removed en bloc by direct extraction through a transvers cavotomy (Figure 1D, 2A). Then, the suprarenal part of the infrahepatic IVC was occluded with a tourniquet and the clamp of intrapericardial IVC was released for the hepatic venous return. A lateral cavectomy around the ostium of the right renal vein was performed. The cavectomy was repaired primarily with a 5.0 polypropylene sutures. The patient was weaned from CPB without any problem. Intraoperative TEE showed no residual IVC tumor. After the heparin neutralization by protamin, a right radical nephrectomy and pulmonary metastasectomy were performed (Figure 2B, C).

The histopathological examination of the resected renal mass revealed that CCSK consisted of cells with fine nuclear chromatin, pale cytoplasm and indistinct cell borders forming nests separated by a fibrovascular stroma. The removed vascular tu-

mor thrombus and excised pulmonary mass showed similar histopathology with renal mass (Figure 2D). The resected margin of ureter, and all lymph node specimens were free of tumor. Immunohistochemically, the cells were diffusely positive for vimentin, CD-117 and negative for Cytokeratin-7, Pancytokeratin, epithelial membrane antigen (EMA), HMB-45, CD-31, Myogenin, Desmin, and Myoglobin. Histopathologic and immunohistochemical features of the tumor were consistent with the diagnosis of CCSK.

The postoperative recovery was uneventful. She was discharged home on the 5th postoperative day in good condition and referred to the oncology unit for chemo-radiotherapy. Adjuvant chemotherapy was started at the 10th postoperative day and she received chemotherapy consisting of doxorubicin, cyclophosphamide, vincristine, and etoposide during 24 weeks period. The patient also received postoperatively, a total of 4950 cGy of radiotherapy in divided doses to the right nephrectomy bed, right lung and IVC.

The patient is now asymptomatic and there was no evidence of local recurrence or distant metastases during the follow-up 18 months after resection.

## DISCUSSION

Although CCSK is uncommon, it is the second most common renal tumor following WT and comprises approximately 5% of all primary renal neoplasms in children. This highly malignant tumor affects children mainly between the ages of 2 and 5 with a male predominance. CCSK is extremely rare in the first 6 months of life and adults [1]. CCSK is often confused with Wilms tumor in children. However, it is classified separately from WT because of several distinctive features. It usually tends to metastasize to bones being different from WT [4].

The common presenting symptoms of these patients include abdominal distension or mass, abdominal pain and hematuria. Approximately 5% of patients have metastatic disease at presentation [5]. CCSK usually tends to metastasize to ipsilateral renal lymph nodes (59%), bones (13%), lung (10%) and liver (9%). CCSK is also able to invade the inferior vena cava with extension into the right atrium [1]. Vascular tumor thrombus is reported in

6% to 9% of cases of WT with an extension to the right atrium noted in 0.7% to 1.1%. Unlike WT, vascular tumor thrombus incidence is almost unknown in CCSK6. However, this is considerably rare and only a limited number of cases of CCSK with vascular tumor thrombus have been reported [1]. There was a complaint of abdominal mass and a lung metastasis as the distant metastasis in our case.

The most intracaval and intraatrial thrombus can be detected by doppler ultrasonography, CECT, and MR angiography. Also, echocardiogram can provide to the additional information on the proximal extent of the intraatrial thrombus (if present) [6]. Although clinical and imaging study characteristics may support a particular diagnosis, histopathologic examination still remains one of the major diagnostic methods.

Grossly, CCSKs usually appeared as a large, well circumscribed, and sharply demarcated unicentric masses ranging from 2.3 to 24 cm in diameter. The tumor arises from the central region in the medulla and replaces normal kidney tissue. Cysts, hemorrhage and small foci of necrosis are commonly present. Microscopically, several histologic variants are recognized. The most common variant is the classic pattern, which is present at least focally in over 90% of tumors. Immunohistochemistry of CCSK consistently shows non-specific positivity for vimentin [1].

Multimodality treatment for CCSK includes the use of chemotherapy and radiotherapy protocols along with surgical approach when possible [7]. After a diagnosis of CCSK is made, radical nephrectomy is the initial treatment of choice if the lesion is resectable [5]. Chemotherapy involves vincristine, cyclophosphamide, doxorubicin and etoposide for stage I to III disease. The patients receive 10.8 Gy of radiotherapy to the flank [1]. Although CCSK is responsive to chemotherapy it is known to have a poor response. Therefore, it becomes imperative to proceed with attempts at complete excision of the approach with its thrombus extension [6]. Various options for the patient of CCSK with vascular tumor thrombus involve tumor balloon thrombectomy, excision via cavotomy, excision via atriotomy, IVC resection with or without graft replacement, and resection under vascular isolation or CPB [6].

CPB with or without hypothermia and cardiac arrest has been recommended for levels III and IV, because it guarantees hemodynamic stability during IVC clamping, an effective control of the intraoperative bleeding and a bloodless field. Therefore, in the absence of effective alternative treatment, complete surgical removal of the primary tumor with its extension along the inferior vena cava is the only suitable cure option. The use of CPB with deep hypothermic circulatory arrest in patients with tumor thrombus extending into the right atrium has been also described. The surgical management of patients with atrial extension (level IV) must be individualized [8]. In our case, nephrectomy with tumor thrombectomy via the IVC and pulmonary metastasectomy were performed with moderate hypothermia CPB (32 °C) without aortic cross-clamping and cardioplegic cardiac arrest.

Despite optimal care, long-term survival is poor and late recurrence is common. Relapses may occur in about 20–40% of the cases. The median time interval to relapse is about 24 months. The overall survival rate is 60-70% [1].

In conclusion, the excision of vascular free floating tumor thrombus extending to the inferior vena cava (level III), renal mass, and lung metastasis associated with CCSK can successfully be performed using CPB without cardiac arrest.

Competing interest: The authors declare that they have no competing interests.

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