—Case Reports—

Successful Transcatheter Arterial Embolization to Control Intratumoral Hemorrhage in Clear-Cell Sarcoma of the Kidney

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Clear-cell sarcoma of the kidney (CCSK) is a rare, aggressive pediatric renal tumor. Intratumoral hemorrhage and tumor rupture are oncologic emergencies requiring a rapid and appropriate response. An 11-year-old boy visited our hospital with abdominal distension of 1 month's duration. Computed tomography (CT) revealed a tumor in the left kidney (size: 200 mm), and analysis of a biopsy specimen confirmed a diagnosis of CCSK. Chemotherapy was initiated to shrink the large, densely vascularized tumor before surgical removal. Two days after starting chemotherapy, the patient developed abdominal and back pain, anemia, and hypotension. CT scanning showed intratumoral bleeding. Emergency transcatheter arterial embolization (TAE) was performed to control the bleeding. Three tumor feeding vessels were identified: an ascending branch from the celiac artery, an intermediate branch from the left renal artery, and a descending branch from the inferior mesenteric artery, of which the intermediate and descending branches were large and bleeding profusely. Therefore, the intermediate branch was injected with ethanol, and the descending branch was treated by gel-foam embolization. Chemotherapy was resumed, and the patient's condition gradually stabilized. The tumor began to shrink, and subsequent chemotherapy progressed well. In week 12 of chemotherapy, the patient underwent tumor resection and left nephrectomy. Postoperative chemotherapy was completed without complications, and there was no recurrence during a 6-year follow-up period. Therefore, TAE can effectively control intratumoral bleeding in pediatric solid tumors, thus preventing high-risk open surgery.
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Key words: transcatheter arterial embolization, clear cell sarcoma of the kidney, hemorrhage, anemia

Introduction

Clear-cell sarcoma of the kidney (CCSK) is a rare pediatric malignant tumor. CCSK accounts for 5% of all primary renal tumors and is mostly observed in children younger than 2 to 3 years[1]. CCSK is rare in Japan: only one to four cases are reported annually[2]. The most common symptoms of CCSK are abdominal masses, abdominal pain, and hematuria. It is an aggressive renal tumor with a worse prognosis than Wilms' tumor, and late recurrence is a concern[1]. Selective embolization of pediatric tumors has been used as an adjunct to chemotherapy and in oncologic emergencies, to reduce tumor size or manage intratumoral hemorrhage preoperatively[3–5]. Although bleeding due to spontaneous tumor rupture is infrequent, it can be fatal when blood loss is massive. This oncologic emergency requires a quick, appropriate response to ensure patient survival.

Here, we report a case of CCSK at an atypical age in a boy with intratumoral hemorrhage, severe abdominal and back pain, and diminished blood pressure. Early transcatheter arterial embolization (TAE) therapy controlled bleeding, prevented tumor rupture, and allowed chemotherapy and surgery to be performed safely at a later time.

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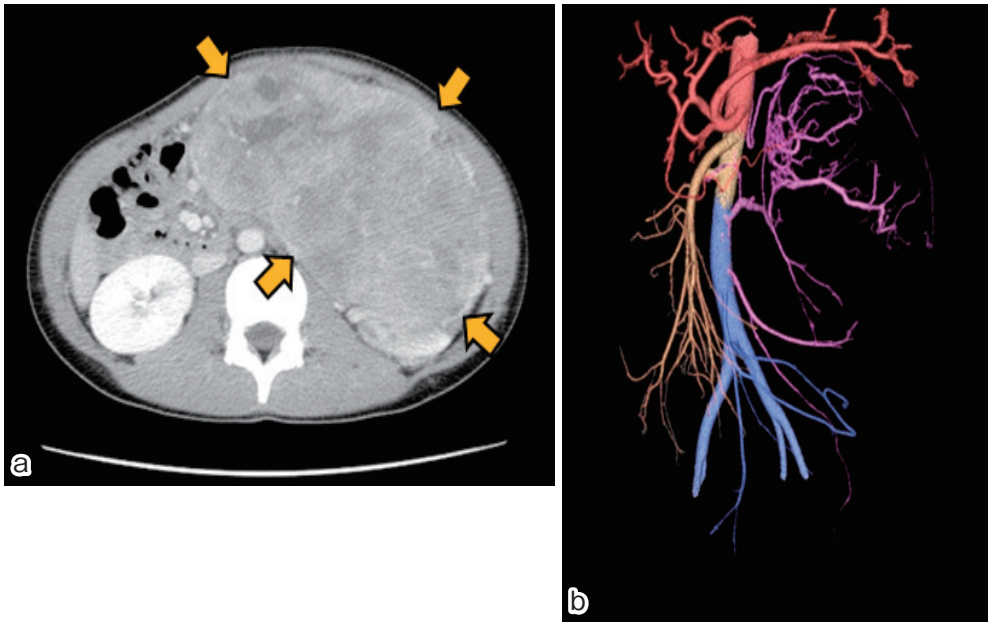

Fig. 1 (a) Contrast-enhanced computed tomography (CT) image at diagnosis. A large tumor in the left kidney is indicated by the arrow. (b) Three-dimensional CT image at diagnosis. The tumor is fed mainly by the ascending branch from the celiac artery, the middle branch from the left renal artery, and the descending branch from the inferior mesenteric artery.

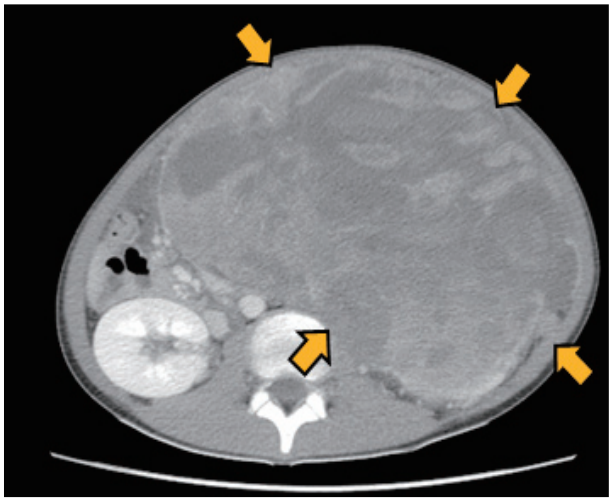

Fig. 2 Contrast-enhanced computed tomography (CT) images after the patient developed sudden abdominal pain and hypotension. The tumor was larger than at diagnosis, suggesting intratumoral bleeding. Extravascular leakage of the contrast agent into the tumor suggested persistent bleeding.

Case Report

An 11-year-old boy presented with a 1-month history of abdominal bloating without dysuria or gastrointestinal symptoms. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a tumor in the left kidney (**Fig. 1a**). CCSK was diagnosed on the basis of findings from an open biopsy and the central review system of the Japan Children's Cancer Group. Bone marrow examination, bone scintigraphy, gallium scan, chest CT, and cranial MRI showed no metastases. The tumor was large, approximately 20 cm along its long axis, and was so densely vascularized that there was a risk of damaging the surrounding large blood vessels intraoperatively (**Fig. 1a, b**). Therefore, operating without preoperative chemotherapy was deemed difficult. A diagnosis of National Wilms' Tumor Study (NWTS) stage III was made, and upfront chemotherapy was started in accordance with the Japan Wilms' Tumor Study-2 protocol regimen I, which is based on the standard NWTS-5 treatment protocol[6].

On the second morning after initial chemotherapy with intravenous vincristine, the patient complained of severe abdominal and lower back pain. His systolic blood pressure decreased, and his hemoglobin level dropped rapidly from 11.8 g/dL on the previous day to 8.2 g/dL. He therefore required multiple red blood cell transfusions. Contrast-enhanced abdominal CT showed tumor enlargement and contrast agent leakage within the tumor (**Fig. 2**). Active intratumoral bleeding was suspected, and emergency TAE was performed to control bleeding, prevent tumor rupture, and stabilize the patient.

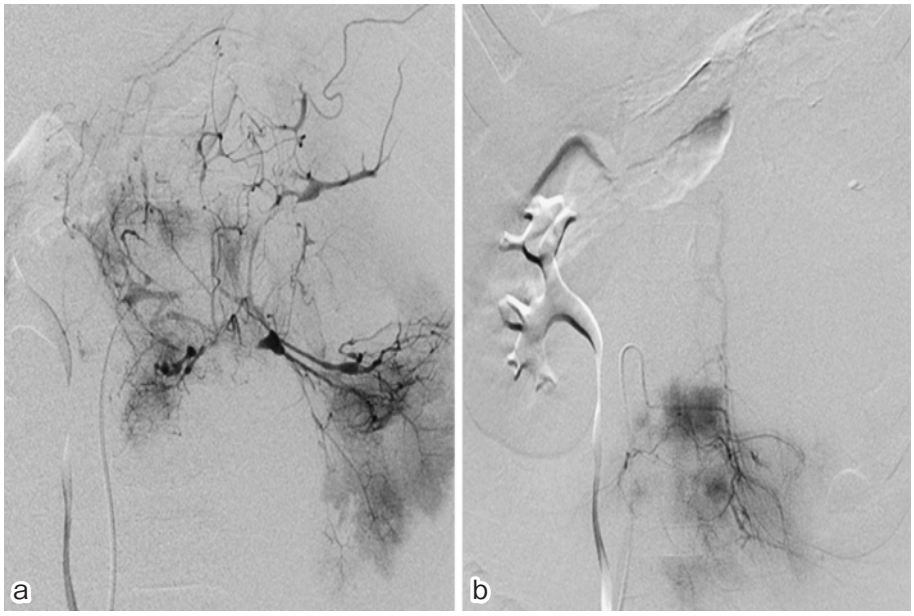

Fig. 3 Angiographic images before transcatheter arterial embolization (TAE). Tumor trophoblastic vessels were found in the (a) ascending branch from the celiac artery and an intermediate branch from the left renal artery, of which the middle branch was thought to have a large amount of bleeding, and (b) a descending branch from the inferior mesenteric artery, which was thought to have a large amount of bleeding.

Three tumor-feeding vessels were identified: an ascending branch from the celiac artery, an intermediate branch from the left renal artery, and a descending branch from the inferior mesenteric artery (**Fig. 3a, b**). Of these, the middle and descending branches were thicker and showed massive bleeding. The middle branch was injected with ethanol, and the descending branch was treated by gel-foam embolization (**Fig. 4a, b**). This helped to control the persistent bleeding. After TAE, abdominal and back pain was alleviated. Signs of tumor lysis syndrome, including elevated uric acid levels, were not observed. However, the patient developed dyspnea due to temporary tumor expansion and circulatory failure, possibly because of intratumoral hemorrhage. Chest radiographs revealed pulmonary congestion. We administered diuretics, albumin replacement, and thrombomodulin α for hypercoagulability. Chemotherapy was resumed 3 days after TAE. Catecholamines were administered during this time. After resuming chemotherapy, the patient's respiratory status improved gradually and the tumor began to shrink, with no further complications. Tumor resection and complicated left nephrectomy were performed in week 12 of chemotherapy (**Fig. 5**). After that, radiation therapy (10.8 Gy) and chemotherapy were completed without complications until week 24, when the last chemotherapy session was conducted. There were no late complications or recurrence during a 6-year follow-up period.

Discussion

According to the Children's Oncology Group high-risk renal tumor study, management of CCSK requires surgery of resectable tumors, followed by administration of vincristine, cyclophosphamide, doxorubicin, and etoposide for stage I to IV disease[6–9]. All patients received postoperative radiation therapy. Patients with CCSK treated according to the NWTS-5 protocol had a 5-year relapse-free survival of 79% and overall survival of 89%. Staging was strongly associated with outcome. The 5-year relapse-free survival rates for stages I, II, III, and IV were 100%, 87%, 74%, and 36%, respectively[8]. The Japan Wilms Tumor Study group developed a protocol that incorporates the NWTS treatment regimen and prioritizes surgery[6]. Preoperative tumor rupture is a major risk for intraperitoneal relapse of the tumor and is thus essential to identify. Although abdominal pain, anemia, and shock suggest tumor rupture, they are nonspecific, and it is difficult to identify tumor rupture on the basis of these symptoms alone[10]. The present patient had a giant kidney tumor on contrast CT scans obtained at the time of hospitalization and required attention because of potential tumor rupture and subsequent bleeding. We should have

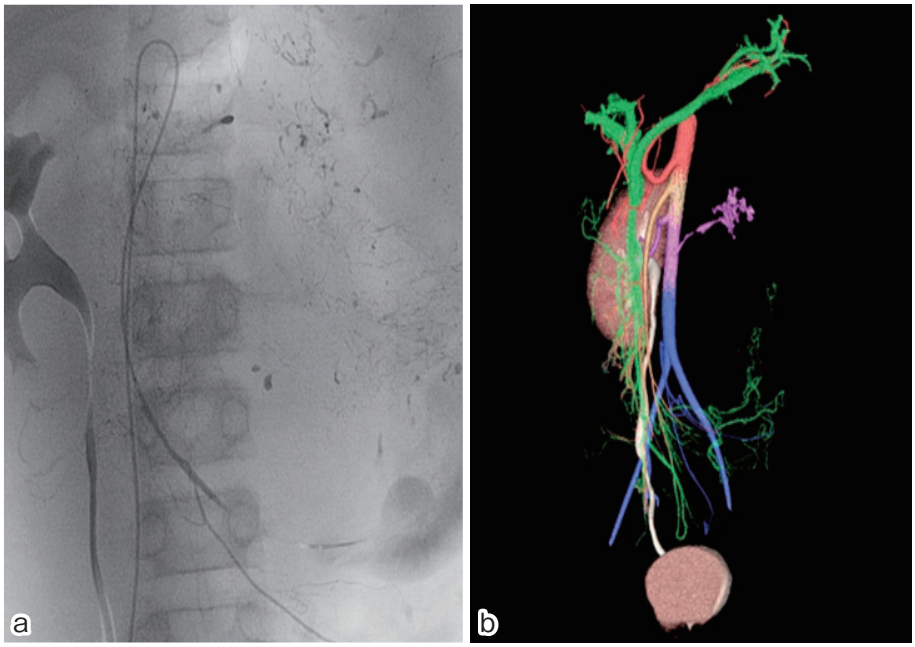

Fig. 4 (a) The middle branch was injected with ethanol. The descending branch was embolized with gel-foam. Hemostasis was confirmed in this angiographic image. (b) After TAE, the middle branch from the left renal artery and the descending branch from the inferior mesenteric artery, which are the trophic vessels of the tumor, cannot be delineated.

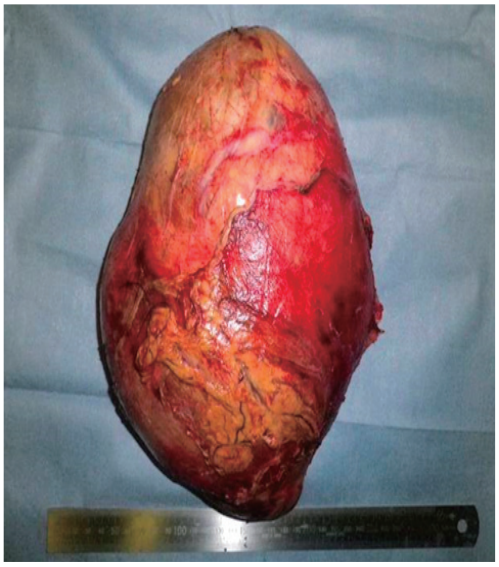

Fig. 5 The tumor (20 cm × 10 cm) was resected.

considered the possibility of intratumor bleeding and tumor rupture.

Khanna et al. reported that CT scans are useful in accurately identifying tumor rupture. CT findings that suggest tumor rupture include 1: tumors with unclear margins; 2: perirenal adipose tissue infiltration; 3: obscured adipose layer around the tumor; 4: retroperitoneal fluid retention; 5: ascites; 6: peritoneal dissemination; 7: ipsilateral pleural effusion; and 8: intratumoral hemorrhage[11]. In particular, the presence of conditions such as an obscured adipose layer around the tumor, retroperitoneal fluid retention, and ascites strongly suggest tumor rupture. The sensitivity of CT scans in detecting tumor rupture is somewhat low, 54 to 80%, but specificity is 88%[11]. In our case, only ipsilateral pleural effusion and intratumoral hemorrhage were present, and it was likely that tumor rupture had not yet occurred.

There are a few reports of children who underwent TAE to manage difficult-to-control bleeding in renal tumors, particularly CCSK. Almgard et al.[12] first performed embolization to treat renal adenocarcinoma in 1973. Since then, renal artery embolization has been increasingly used to treat advanced or unresectable renal tumors in adults. Clinical studies of adults have shown that preoperative renal embolization significantly reduces blood loss during nephrectomy, especially in large hypervascular tumors[13,14]. Renal artery embolization has also been used to manage malignant renal tumors in children, mainly in those with Wilms' tumor[15,16]. Chitnis et al.[16] reported that, although therapeutic embolization is not routinely performed for pediatric solid tumors, TAE may be indicated when a patient's general condition is poor and they may not be able to withstand surgery. Smith reported that patients rarely fail to respond to blood trans-

fusion for severe bleeding from tumors; however, such tumors can be fatal and TAE may be effective in such cases[3].

The present patient had progressive anemia despite multiple blood transfusions and severe symptoms. We therefore decided that TAE would be a more effective, less invasive option than immediate excisional surgery to control bleeding. Accordingly, tumor rupture was prevented by rapid TAE. After TAE, the patient's general condition improved to a level where uncomplicated chemotherapy, and nephrectomy and radiotherapy at a later date, was feasible. TAE is an effective tool for controlling intratumoral bleeding in pediatric solid tumors.

Conclusion

A rare pediatric renal tumor, CCSK, complicated by intratumoral hemorrhage during chemotherapy was successfully managed by TAE. Use of TAE obviated high-risk open surgery and enabled the planned chemotherapy to be effectively administered in conjunction with a later successful and less risky tumor removal procedure.

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Conflict of Interest: The authors declare no conflict of interest.

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