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Pediatric clear cell sarcoma of the kidney with cavoatrial thrombus

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Abstract Clear cell sarcoma of the kidney (CCSK) is a rare renal tumor. Only 4 cases of CCSK with vascular thrombus have been reported, and 2 of these were pediatric cases. One of the children had an intraatrial thrombus as well. We describe a 3-year-old boy who was diagnosed as having a Wilms tumor but did not respond to preresection chemotherapy. He underwent complete resection of the tumor under cardiopulmonary bypass. Histologic examination indicated that the tumor was a CCSK. The patient was then managed with appropriate chemotherapy and radiation therapy and is well 16 months after diagnosis.

Clear cell sarcoma of the kidney (CCSK) is a rare renal tumor comprising approximately 3% of all renal tumors in children [1,2]. Unlike Wilms tumor, vascular thrombus is almost unknown in CCSK. Only 4 cases of CCSK with vascular thrombus have been previously reported, and of these, only 2 were in the pediatric age group [3-6]. Thrombus extending to right atrium is an uncommon finding in Wilms tumor with an incidence of 0.7% to 1.1% [2,6]. It is rare in CCSK, with only 3 such reported cases [3-6]. Herein we report a case of a 3-year-old boy with CCSK with vascular thrombus extending to the right atrium that was managed successfully by excision of the mass and thrombus under cardiopulmonary bypass.

1. Case history

A 3-year-old boy presented with a gradually progressive right-sided abdominal mass of 7-month duration. He was investigated at another center with a contrast-enhanced computed tomographic (CT) scan (CECT) of the abdomen and chest, which showed a right-sided solid renal tumor, with vascular thrombus extending to the right atrium (Fig. 1A-C). Fine-needle aspiration cytology was suggestive of Wilms tumor; hence, neoadjuvant chemotherapy consisting of vincristine, dactinomycin, and doxorubicin was started. However, after 8 weeks of standard chemotherapy, a repeat CECT of the abdomen showed no reduction in size of the renal tumor or the thrombus. It was then that the child was referred to our care. At transfer, the child had a 15 × 12-cm firm, nontender right-sided renal mass. The presence and the extent of the vascular thrombus were confirmed by an

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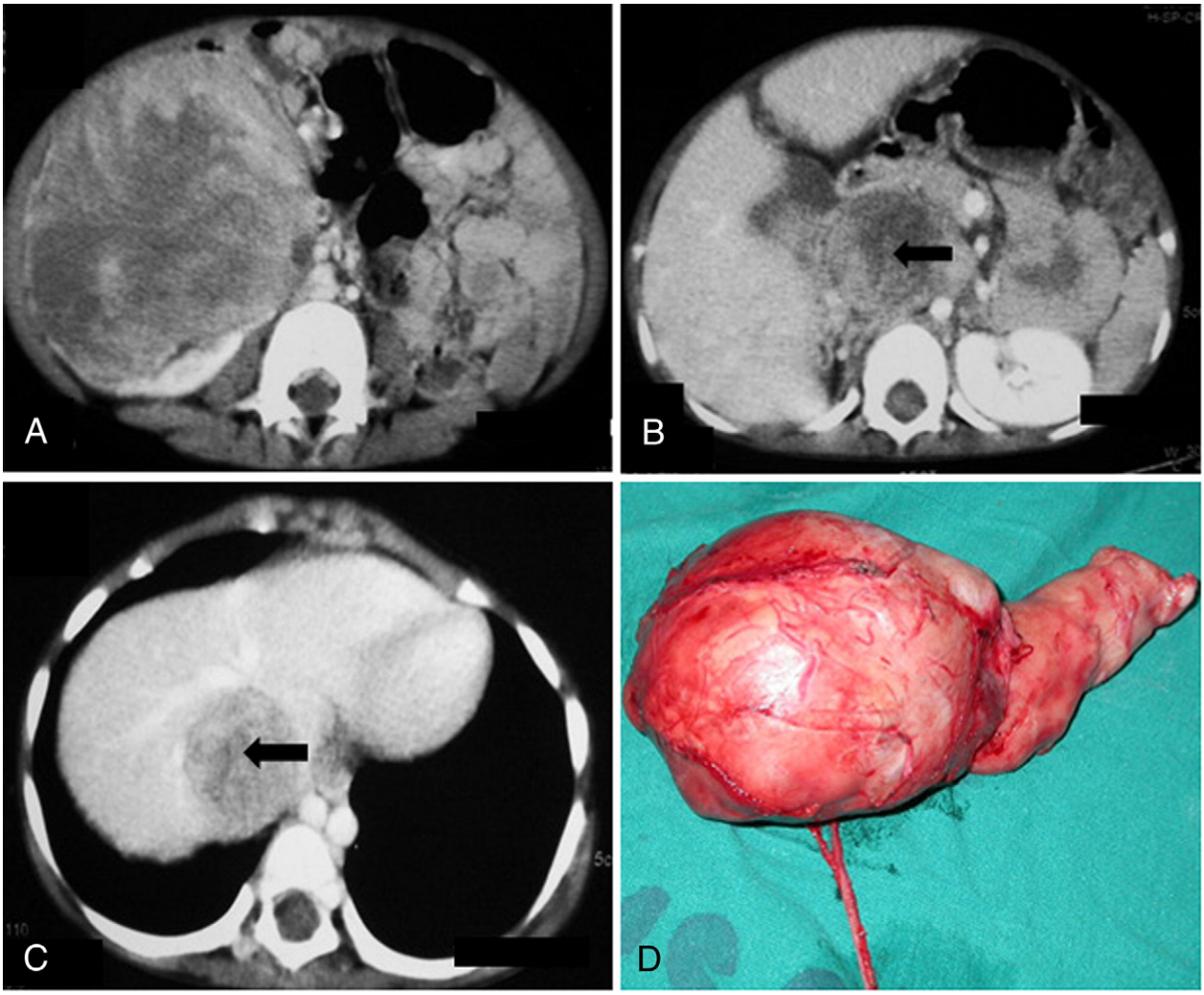

Fig. 1 Contrast-enhanced CT scan at initial presentation showing a large right renal mass (A), thrombus extending into the retrohepatic IVC (B; arrow), thrombus extension into the right atrium (C; arrow), and gross specimen of the resected kidney with the caval and atrial thrombus (D).

ultrasound Doppler study. A 2-dimensional echocardiogram revealed an inferior vena cava (IVC) thrombus with a 25 × 35-mm thrombus extending from suprahepatic IVC into the atrium. No superior vena caval obstruction, vegetation, or pulmonary embolism was found, and normal biventricular function was reported.

The child was explored through a median sternotomy incision joining a roof top-right subcostal incision that extended across the midline. A transesophageal Doppler probe was inserted to ascertain the proximal extent of the tumor thrombus intraoperatively and also to detect a patent foramen ovale if present. The child was subsequently put on normothermic cardiopulmonary bypass, without arresting the heart, through the aorta, superior vena cava, and right common femoral vein (Fig. 2). The pulmonary artery trunk was controlled to prevent tumor embolization into the lungs. After division of the falciform ligament and the right triangular ligaments of the liver, the right lobe of the liver was retracted medially to expose the entire length of the infradiaphragmatic IVC. The right renal mass was mobilized, and the right renal artery and ureter were divided between ligatures. Control of the left renal vein and the IVC was obtained just proximal to the renal veins. The right renal vein was then opened, between stay sutures, in continuity with cavotomy, which was extended superiorly onto the right atrium. The entire thrombus was then removed en bloc with the renal mass using endarterectomy instruments (Fig. 1 D). Part of the thrombus entering the hepatic veins was also removed. Paracaval and paraaortic lymph node sampling was done as per the protocol for Wilms tumor. The IVC and right atrium were repaired primarily with polypropylene sutures. The patient was then weaned from cardiopulmonary bypass, and the wounds closed.

The histopathology of the resected renal mass and the thrombus revealed a tumor with polygonal cells and moderate cytoplasm with spindling and palisading of nuclei and arborizing blood vessels. There were areas of cystic change and focal areas of necrosis. There was no breach of the renal

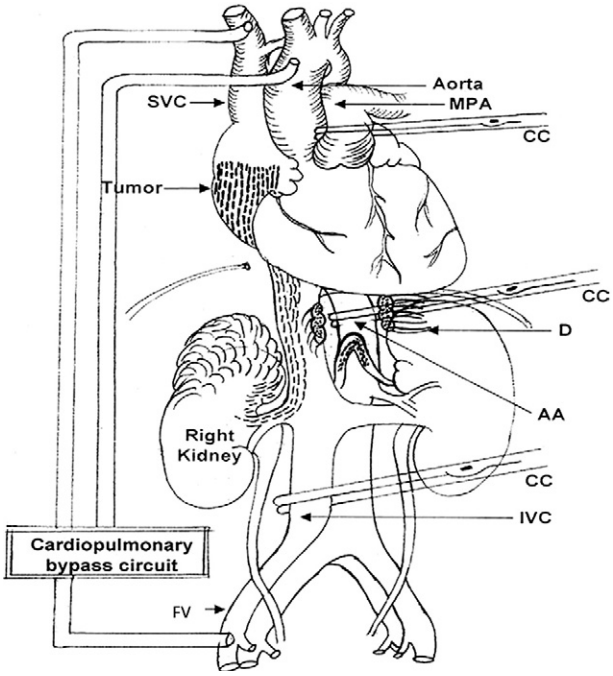

Fig. 2 Schematic diagram for the cardiopulmonary circuit used. SVC indicates superior vena cava; MPA, main pulmonary artery; CC, cross clamp; AA, abdominal aorta; D, diaphragm; FV, femoral vein.

capsule, and the hilar vessels, the resected margin of ureter, and all lymph node specimens were free of tumor. The tumor thrombus showed similar histology with focal areas of necrosis and fibrosis. Histopathology was consistent with CCSK. Because bone and brain metastasis are common in CCSK, a postoperative CT scan of the head, skeletal survey, and bone scans were obtained and were negative for brain or bone metastases. The patient was staged as stage 3 (National Wilms Tumor Study staging) in view of the adherent vascular thrombus and the preresection chemotherapy received. The child received 24 weeks of chemotherapy for CCSK, consisting of doxorubicin, cyclophosphamide, vincristine, and etoposide (regimen I: National Wilms Tumor Study-5 protocol). He also received radiotherapy to the flank (1080 cGy in 7 fractions). The patient was followed every 3 months for the first year and underwent surveillance imaging for any local or distant metastases and will be seen in 6-monthly follow-up for the next 5 years, considering the propensity for late metastatic recurrence in CCSK. Currently, the child has completed all treatment and is disease free at 16 months from diagnosis. The present imaging investigations do not reveal any local or distant recurrence. The Doppler studies do not show any flow in the retrohepatic portion of the IVC; however, the patient does not have any pedal edema or hepatomegaly.

2. Discussion

Vascular thrombus is reported in 6% to 9% of cases of Wilms tumor with an intraatrial extension noted in 0.7% to 1.1% [2,7,12]. Only 4 cases of CCSK with vascular thrombus have been reported in the English literature, and only 2 in the pediatric age group [5,6]. Of these 4 cases, 3 had tumor thrombus extending into the atrium [3-5]. Most intracaval and intraatrial thrombus can be delineated by CECT and a Doppler study. However, a magnetic resonance imaging with magnetic resonance imaging angiogram will be able to profile the thrombus more accurately. An echocardiogram will also add to the information on the extent of the intracardiac thrombus, the structure of the heart, and the ventricular function. Most intracaval and intraatrial thrombi in Wilms tumor show a response to chemotherapy, and either resolves completely or can be removed through a cavotomy. Very few cases require exploration under cardiopulmonary bypass [7]. There are variants of Wilms tumor that do not respond rapidly to chemotherapy. If after 8 weeks of chemotherapy, the tumor still remains in the atria, it should be resected instead of persisting with more chemotherapy. This is the same practice used in Wilms tumor because preoperative histologic or cytologic diagnosis of CCSK is not always available. On histologic examination, the diagnosis of CCSK may be missed if entrapped tubules are misinterpreted because tubular differentiation or spindled areas are misinterpreted as mesenchymal differentiation in Wilms tumor [8]. Clear cell sarcoma of the kidney is responsive to chemotherapy but 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. A multidisciplinary approach to treatment involving the pediatric surgeons, cardiac surgeons, anesthesiologists, cardiologists, radiologists, medical oncologists, and radiation oncologists is advised in such cases. Various options include tumor balloon thrombectomy, excision via atriotomy, IVC resection with or without graft replacement, and resection under vascular isolation or cardiopulmonary bypass [2,7,9-12]. The operative approach in such cases demands attention toward certain specific aspects of the procedure. Because friability of the atrial thrombus, there is a risk of pulmonary or even arterial embolization (through patent foramen ovale if present). To prevent this careful handling and control of the pulmonary artery before any tumor manipulation is an important technical consideration. The IVC is obviously not appropriate for use in cardiopulmonary bypass in such cases; therefore, bypass needs to be established via alternative veins. In our case, the right common femoral vein was used. After having received some preresection chemotherapy, there is a possibility of the tumor thrombus being adhered to the IVC wall, and one may require excising the segment of IVC followed by the use of a graft to restore the continuity as has been observed in Wilms tumors. In the present case, as well as the one reported previously [5], fortunately, the thrombus could be removed using endarterectomy techniques, and the IVC and atrium could be repaired primarily without requiring a graft or patch interposition. Including the present case, 4 of 5 cases of CCSK with vascular thrombus

had atrial extension [3-5]. Whether atrial extension and this kind of nonadherent thrombus are characteristics of CCSK is something that may become clearer with additional experience in future cases.

This rare case of CCSK with cavoatrial thrombus demonstrates that not all cases of renal tumors with vascular thrombus in children are Wilms tumor. Alternate histologic diagnosis should be considered especially in cases of renal tumors where the vascular thrombus does not show any response to preoperative chemotherapy. Considering the fact that CCSK has a poor response to chemotherapy, complete excision of the tumor and thrombus should be attempted, even if it entails extensive surgery under cardiopulmonary bypass.

References

[1] Argani P, Perlman EJ, Breslow NE, et al. Clear cell sarcoma of the kidney: a review of 351 cases from the National Wilms Tumor study Group pathology center. Am J Surg Pathol 2000;24:4-18.

[2] Shamberger RC, Ritchey ML, Haase GM, et al. Intravascular extension of Wilms tumor. Ann Surg 2001;234:116-21.

[3] Bhayani SB, Liapis H, Kibel AS. Adult clear cell sarcoma of the kidney with atrial tumor thrombus. J Urol 2001;165:896-7.

[4] Toyoda Y, Yamashita C, Sugimoto T, et al. Clear cell sarcoma of the kidney with tumor extension into the right atrium. J CardiovascSurg (Torino) 1998;39:489-91.

[5] Zigman A, Shen I. Clear cell sarcoma of the kidney with cavo-atrial tumor thrombus: complete resection in a child. J Pediatr Surg 2006;41:1464-6.

[6] Ohtake N, Shiono A, Okabe K, et al. Clear cell sarcoma extending into inferior vena cava. Nippon Hinyokika Gakkai Zasshi 1995;86:1298-301.

[7] Ritchey ML, Kelalis PP, Breslow N, et al. Intracaval and atrial involvement with nephroblastoma: review of National Wilms tumor Study-3. J Urol 1988;140:1113-8.

[8] Iyer VK, Agarwala S, Verma K. Fine needle aspiration cytology of clear cell sarcoma of the kidney: a study of eight cases. Diagn Cytopathol 2005;33:83-9.

[9] Nakayama DK, Norkool P, deLorimier AA, et al. Intracardiac extension of Wilms' tumor. A report of the national Wilms' tumor study. Ann Surg 1986;204:693-7.

[10] Thompson WR, Newman K, Seibel N, et al. A strategy for resection of Wilms' tumor with vena cava or atrial extension. J PediatrSurg 1992; 27:912-5.

[11] Lodge AJ, Jaggers J, Adams D, et al. Vascular control for resection of suprahepatic intracaval Wilms' tumor: technical considerations. J Pediatr Surg 2000;35:1836-7.

[12] Luck SR, DeLeon S, Shkolnik A, et al. Intracardiac Wilms' tumor diagnosis and management. J Pediatr Surg 1982;17:551-4.