Cavectomy for the Treatment of Wilms Tumor With Vascular Extension

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Purpose: Vascular extension to the vena cava occurs in 4% of Wilms tumor cases and can reach the right atrium in up to 1%. When this happens the thrombus is usually not adherent to the vessel wall, and there is blood flow around it. Preoperative chemotherapy can cause thrombus regression and even resolution. If the thrombus persists after chemotherapy, surgery will be a challenge. On the other hand, if the thrombus invades the vessel wall, its removal may not be feasible. In this situation cavectomy is a good surgical strategy because it provides complete resection. The prerequisite for cavectomy is the absence of blood flow in the vena cava on preoperative Doppler ultrasonography. We report 3 cases of Wilms tumor with vena caval invasion in which cavectomy was performed, and discuss the principles, indications and operative technique.

Materials and Methods: A total of 171 patients with Wilms tumor were treated at our institution between 1984 and 2004. Of these patients 6 with intravascular extension of thrombus within the right atrium were treated with extracorporeal circulation, cardiac arrest and profound hypothermia, and 3 were treated with cavectomy.

Results: There were no instances of surgical complications or postoperative renal failure in our patients who underwent cavectomy. All remain well and free of disease.

Conclusions: Cavectomy is a safe procedure for treating pediatric patients with Wilms tumor when there is extension and invasion of the vena cava wall without blood flow.

Key Words: Wilms tumor; vena cava, inferior; venous thrombosis; kidney neoplasms

Wilms tumor is the most frequent renal tumor in childhood, corresponding to approximately 90% of kidney neoplasms in pediatric patients. Vascular extension to the vena cava occurs in 4% of the cases and can reach the right atrium in 0.8% to 1%.[1–3] When this happens the vessel wall is usually not invaded by the tumor thrombus and there is blood flow around it.

Preoperative chemotherapy can cause thrombus regression and even resolution.[2,4,5] If the thrombus persists after PC, surgery will be a challenge, and in some cases extracorporeal circulation is needed. With this technique the thrombus can be removed by an incision in the vena cava, since there is no invasion of the vessel wall. On the other hand, if the thrombus invades or is firmly adhered to the vessel wall, its removal may not be possible. In this situation cavectomy is a good surgical strategy for complete resection of the tumor. The precise indication for cavectomy is the absence of blood flow in the vena cava. Little is reported in the literature about this procedure in children with WT. We report 3 cases of WT with vascular invasion in which cavectomy was performed.

PATIENTS AND METHODS

We performed a retrospective review of 171 patients with Wilms tumor treated at our institution between 1984 and 2004. In 3 of these patients cavectomy was chosen to achieve complete resection of the tumor.

Case 1

VRS, a 4-year-old girl, presented with hematuria and hypertension. Ultrasound showed a 6.8 × 6.4 cm right renal mass, with vascular extension to the vena cava from the renal veins to a distance of 3 cm from the right atrium. Excretory urography revealed right renal exclusion. Preoperative CT and cavography demonstrated vena caval obstruction and venous return through collateral veins (fig. 1).

The patient received 7 cycles of PC consisting of vincristine, doxorubicin and dactinomycin. Another cavography was performed and again showed caval obstruction. The intraoperative finding of an adherent tumor thrombus invading the vena cava wall indicated cavectomy. "En bloc" nephrectomy and cavectomy were performed. The resection of the vena cava extended from the iliac veins to its retrohepatic portion. The patient did well and has normal renal function.

Pathological evaluation revealed Wilms tumor with capsular invasion. The renal vein and vena cava had organized neoplastic thrombosis. The case was considered stage III. Treatment consisted of 15 months of chemotherapy accompanied by abdominal radiotherapy. The patient was in remission at 11 years postoperatively.

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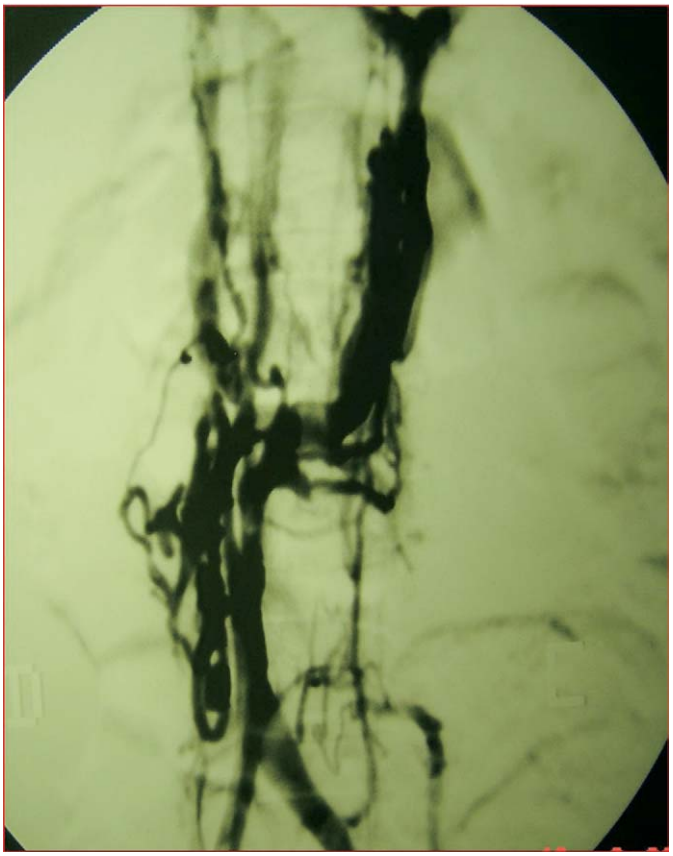

FIG. 1. Cavography shows vena caval obstruction and collateral vessels.

Case 2

BBS, a 3-year-old girl, presented in 2004 with a 3-month history of an abdominal mass with rapid growth in the prior week, following a fall. Ultrasound demonstrated a huge renal mass in the right kidney, and Doppler showed a thrombus from the iliac bifurcation to the hepatic veins. Computerized tomography revealed the same tumor and renal exclusion (fig. 2).

FIG. 2. Computerized tomography reveals huge mass in right kidney (case 2).

FIG. 3. Computerized tomography demonstrates reduction of renal mass.

The patient received 6 cycles of chemotherapy according the SIOP protocol. Shrinkage of the renal mass was observed, although the thrombus persisted to the same extent, as did the absence of blood flow in the vena cava (fig. 3). Echocardiography demonstrated no mass in the atrium. Laparotomy was performed, and the vena cava was found to have a thrombus invading its wall as well as an absence of blood flow. "En bloc" resection was performed, including the right kidney and the vena cava from the iliac bifurcation to its retrohepatic portion (fig. 4). The left renal vein was ligated next to the vena cava, and as in the first case, there was no need for extracorporeal circulation. The patient did well and was discharged from the hospital at 7 days postoperatively with normal renal function.

Pathological evaluation showed total necrosis of the tumor. Chemotherapy was completed. Recovery was uneventful and the patient was free of disease at 1.5 years postoperatively.

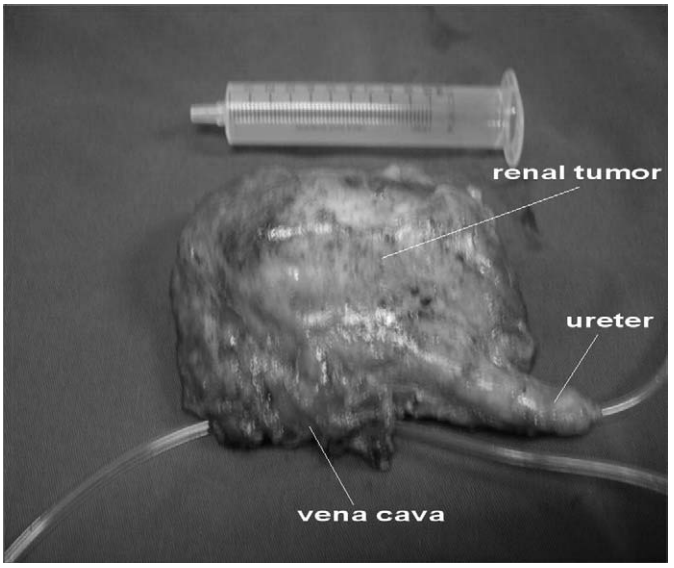

FIG. 4. En bloc resection of right kidney with vena cava