Nephron Sparing Surgery for Synchronous Bilateral Wilms—Operative Steps

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Abstract

Approximately 5% of children with Wilms tumor present with bilateral disease. The treatment challenge is to achieve a high cure rate while maintaining adequate long-term renal function. We report the feasibility and outcome of nephron sparing surgery in a child with bilateral Wilms tumor who was treated at our institution.

Keywords Bilateral Wilms tumor · Nephron sparing surgery · Postoperative complications · Renal function

Introduction

Wilms tumor is the most common renal tumor in children. It affects approximately 650 children in the USA each year. Approximately 5% of these will have synchronous bilateral disease [1] and 2–3% will develop a contralateral metachronous tumor [2]. Bilateral Wilms tumor (BWT) is more commonly associated with genetic syndromes which often include intrinsic renal disease, and is known to present earlier [1]. The most significant morbidity associated with BWT is renal insufficiency and most often results due to resection of a large portion of the renal parenchyma. End-stage renal disease (ESRD) adversely affects both the overall health of the child as well as the quality of life. These effects are most pronounced in young children. Any procedure that preserves native renal function for a prolonged period of time following surgery carries a clear theoretical benefit. Nephron sparing surgery (NSS) in recent years has been become the standard

of care for the management of most adult renal tumors (T1a, T1b and T2) with excellent oncologic control and functional outcomes. Due to the rarity of BWT, NSS has been less commonly used in pediatric renal tumors, but has been shown to provide excellent oncologic control and at the same time preserve renal function [3, 4]. In this article, we describe the operative steps of NSS in a child with BWT.

Case Report

A 2-year-old male child presented to the pediatric urological services of the hospital with complaints of distension of the abdomen. The child had normal mile stones and was vaccinated as per the schedule. The child was anemic (Hb—9.2 mg%) and weighed 10.5 kg. Serum creatinine was 0.24 mg%. Ultrasonographic examination revealed bilateral renal masses. Computed tomography (CT) showed a heterogeneously enhancing lobulated mass $2.8 \times 3.1 \times 4.2$ cm in the upper pole of the right kidney and a hypo-enhancing solid mass $6 \times 6 \times 7$ cm upper and mid polar mass in the left kidney. No other lesion was identified. Chest x-ray was normal.

A diagnosis of bilateral Wilms tumor was made and the child was put on preoperative chemotherapy (SIOP-2001 guidelines) consisting of vincristine injection 0.75 mg on days 1, 8, 15, 22, 29, and 36, and actinomycin D injection 475 mcg on days 1, 15, and 29. The child was monitored with serial blood tests consisting of total WBC counts, differential WBC count, liver function tests, hemoglobin, and electrolytes. There was massive reduction (> 50%) in the size of the renal masses bilaterally as seen on CT (Fig. 1a, b).

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Fig. 1 **a** Prechemotherapy CT scan shows bilateral huge tumors, occupying more than 50% of the kidney. **b** Post-chemotherapy the size of the tumors are reduced to more than 50%

Surgical Approach

It was decided to perform bilateral nephron sparing procedures during a single operation so as to remove all macroscopic tumor at the earliest feasible time so that the chance of developing therapy-resistant disease was minimized and spare the child from an additional operation, thereby minimizing interruption of chemotherapy. The operation planned was similar to that described by Cozzi and Zani.[5].

The peritoneal cavity was entered through a transverse upper abdominal incision (Fig. 2a). The kidney with the larger tumor burden was approached first, was mobilized completely, and was elevated on its vascular pedicle with a vessel loupe placed around the ureter. The lesions to be excised were identified, and the capsule of the kidney was scored with electrocautery to outline the planned extent of resection (Fig. 2b). Whenever possible, a small rim of normal kidney (0.5–1 cm) was included around the resected lesion. The resection proceeded generally within a well-defined plain between the

tumor and kidney (Fig. 3a). Large intraparenchymal vessels were controlled with absorbable ligatures. Hemostasis along the parenchymal surface was then completed with argon beam coagulation. Temporary vascular occlusion was used by clamping the renal vessels. Careful handling of the vascular pedicle remains a critically important step, as traction injuries with vascular thrombosis can occur, especially in children.

Care was taken to determine whether the collecting system was violated or entered, which is known to occur commonly. In our case, no such breach was identified. In case such a breach is identified, the cut edges of the violated calyx are closed with absorbable suture, often after placing a “double-J” ureteral stent. The kidney was folded over the cut surface, encasing a piece of Surgicel, to assure additional hemostasis and that the kidney retains a more reniform contour (Fig. 3b). A similar approach was used for the contralateral kidney (Fig. 4a, b). Removal of regional lymph nodes was performed following NSS. Flank suction drains were placed. The abdomen was closed in layers after ensuring hemostasis. The

Fig. 2 **a** Transverse upper abdominal incision. **b** The capsule of the kidney scored with electric cautery to outline the planned extent of resection

Fig. 3 **a** The excision is proceeded within a well-defined plain between the tumor and the kidney. **b** The kidney is folded over a piece of Surgicel or bolster and the two cut ends sutured over the bolster

Penrose drains were removed on the third postoperative day and urethral catheter after 48 h. The child was not given any blood transfusions either intraoperatively or postoperatively.

Serum creatinine estimated on the fourth postoperative day was 0.42 mg% and serum hemoglobin was 12.5 mg%. The child had a smooth recovery postoperatively. The child was initiated on further chemotherapy on day 10 in the postoperative period.

Discussion

The survival for patients with Wilms tumor has improved due to the use of more effective, multimodality therapy, and this has led to decrease in the intensity of therapy in order to avoid therapy-related complications [2]. This includes the extent of surgical resection also. Several authors have adopted a surgical approach in these patients with BWT, wherein preoperative chemotherapy is followed by nephron sparing procedures on both the kidneys as the initial, definitive surgical procedure for all children, regardless of the radiographic appearance of the tumors and kidneys [6]. Despite this aggressive surgical approach, the results with regard to complications and to functional and oncologic outcomes compare favorably with those obtained through the larger cooperative groups (Children's Oncology Group, International Society of Pediatric Oncology) [7].

The management of BWT has evolved from primary surgical extirpation to kidney-preserving resection after preoperative chemotherapy. Preoperative chemotherapy often results in significant reduction in tumor size, thereby facilitating subsequent renal salvage. Extended follow-up of patients with BWT who were enrolled on NWTS-2 and -3 showed no difference in survival rates between those treated initially with surgical resection and those treated with biopsy and preoperative chemotherapy [8]. The NWTS-5 recommendation for the management of bilateral WT includes initial biopsy and local staging followed by chemotherapy (according to abdominal stage and histologic features) and second-look surgery at week 5. If needed, additional chemotherapy or radiation therapy is given, but definitive surgery is recommended within 12 weeks of diagnosis to limit the risk of chemoresistant clonal expansion [9].

According to the SIOP-93 guidelines, preoperative chemotherapy is recommended in BWT. Treatment is continued as long as there is imaging evidence of tumor regression. When regression is not sufficient to perform nephron sparing surgery, other drugs are to be used. Bilateral or at least unilateral nephron sparing surgery is performed when technically possible. Surgery is performed in one step (bilateral surgery at the same time) or two steps to improve patient tolerance, or by a few weeks (> 1 month) to allow for the administration of chemotherapy matched to the type and stage of the resected tumor. Postsurgical management is chemotherapy, adjusted to

Fig. 4 **a** Excision of the tumor on the contralateral kidney. **b** Excised specimen from both the kidneys

fit the highest histological grade and local stage of the tumor. In patients with stage III disease, complementary radiotherapy is delivered to a maximum dose of 12 Gy (11) [10].

The focus of treatment in children with BWT is survival, and the preservation of good long-term renal function. Nephron sparing surgery for children with bilateral Wilms tumors is nearly always technically feasible, with acceptable complications. This approach, when combined with adjuvant therapy, provides an opportunity to preserve renal function while maintaining a high probability of cure and should be considered strongly for all patients who have synchronous, bilateral Wilms tumors, despite the radiographic appearance of the kidneys and associated tumors.

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