Urology Case Reports 18 (2018) 16–18

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Extensive synchronous bilateral Wilms tumor treated with nephron sparing surgery

Laura DiChiacchio [a, *], Nicole M. Shockcor [a], Regina Macatangay [b], Eric Strauch [a]

[a] University of Maryland Medical Center, Department of Surgery, USA
[b] University of Maryland Medical Center, Department of Pediatrics, USA

ARTICLE INFO

Article history:
Received 16 January 2018
Received in revised form
6 February 2018
Accepted 14 February 2018
Available online 21 March 2018

Introduction

Wilms tumor presents with 650 cases annually in the U.S. alone; 5% of these cases present as simultaneous bilateral Wilms tumor while an additional 2–3% present with contralateral metachronous disease.[1] Bilateral Wilms tumor (BWT) in particular is associated with an increased risk of long-term renal insufficiency, 10% as compared to 0.7% in unilateral Wilms tumor.[1] BWT is also associated with genetic syndromes such as Beckwith-Wiedemann, Denys-Drash, and Wilms-aniridia-genitourinary-mental retardation (WAGR) among others. Standard treatment of BWT is based on NWTS 4 Regimen DD-4 with 12 weeks of neo-adjuvant chemotherapy followed by definitive surgical resection; The Children's Oncology Group protocol (AREN 0534) stipulates the importance of obtaining negative margins with lymph node sampling including paraaortic and pericaval nodes.[2] Pre-operative imaging is essential for surgical planning, and generally underestimates the extent of healthy parenchyma given compression of neighboring oncologic lesions. Use of intra-operative ultrasound allows more accurate delineation of areas requiring resection, particularly in extensive NSS as in the case below. This report contributes to the body of literature reporting equivalent long-term overall and disease-free survival with NSS compared to unilateral partial nephrectomy with contralateral radical nephrectomy in BWT. Improved long-term negative sequelae including hypertension is also described in this case, despite extensive bilateral disease.

Case report

A 2-year-old boy with a history of chronic constipation was seen by his primary care physician after an abdominal mass was felt. An ultrasound and subsequent CT abdomen and pelvis was obtained demonstrating multiple renal masses suspicious for multifocal Wilms tumor with evidence of IVC compression and left hydronephrosis. A CT chest showed no evidence of metastases.

An MRI abdomen and pelvis was performed with the largest left renal lesion in the upper pole measuring 4.6 × 4.7 × 3.3 cm (width, craniocaudal, anteroposterior) and hyperintense on T2 imaging with heterogeneous enhancement. The largest right renal mass measured 8.1 × 7x5.7 cm (anteroposterior, craniocaudal, width) and was located in the inferomedial pole. An exact number of lesions in each kidney could not be obtained due to the size and number of the lesions, but at least 10 separate lesions were identified in each kidney. Two days after his initial complaint, a left subclavian 6.6 Fr Broviac was placed for chemotherapy access and he was started on Children's Oncology Group AREN 0534 (vincristine, actinomycin, and doxorubicin). Further preoperative work up was completed with a MAG 3 (radioisotope renography) study describing 53% function of the left kidney and 47% of the right kidney; a renal duplex illustrated minimal diameter reduction to bilateral renal arteries. Prior to surgical intervention, he was prescribed two antihypertensives for hypertension >95% for age. After 12 weeks of chemotherapy, a repeat MRI abdomen and pelvis showed significant decrease in disease burden (Figs. 1 and 2). A renal sparing bilateral resection was performed with the removal of nine tumors from the right and five from the left. The left superior renal hilar lesion was found to be adherent to the left main renal artery and vein. This was peeled off and determined to have a negative margin. No lymph node (0/5) involvement was described on final pathology. As the patient was stage 3 post resection with inadequate margins, 12 additional weeks of chemotherapy were performed as well as bilateral flank radiation (total 10.5 Gy in 7 fractions). Three years post chemo, he shows no signs of recurrent disease or evidence of metastases (Fig. 3). He continues to require

* Corresponding author.
E-mail address: ldichiacchio@som.umaryland.edu (L. DiChiacchio).

https://doi.org/10.1016/j.eucr.2018.02.008

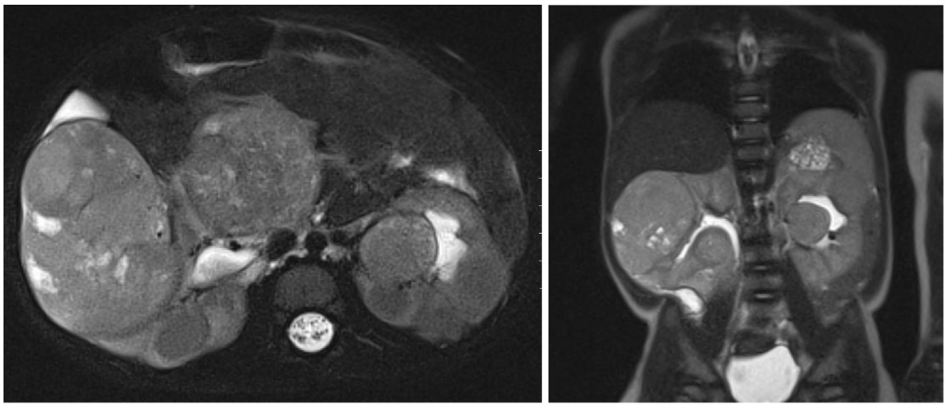

Fig. 1. Representative images from MRI abdomen obtained upon initial presentation. Multiple large bilateral masses; left upper pole mass measuring 4.6 × 4.2 × 3.3cm and right inferomedial mass measuring 7.4 × 6.1 × 6.6cm both apparent in crosssection here.

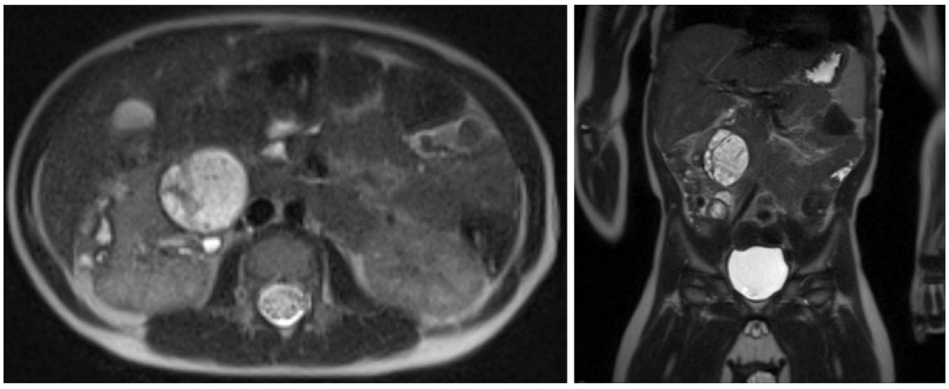

Fig. 2. Representative slices from MRI abdomen after 11 cycles of chemotherapy (COG AREN 0534). Multiple bilateral masses mainly stable to slightly decreased in size, largest mass (right) decreased from 7.4 × 6.1 × 6.6cm to 4.2 × 3.5 × 3.0cm.

no antihypertensives and his baseline creatinine remains 0.35–0.5 mg/dL.

Discussion

In a retrospective review of 42 patients presenting with BWT to a single institution between 2001 and 2014, 39 underwent NSS. At mean follow-up of 4.1 years none of these patients were diagnosed with ESRD, with all eGFRs >60mL/min/1.73m2 at last follow-up.[3] Contemporary studies reported in 2010 or later comparing radical nephrectomy (RN) and NSS universally reported equivalent responses; however, these studies were limited by small numbers in addition to inconsistent reporting and variable operative technique.[4] Post-surgical outcomes typically reported include renal function, hypertension, and presence or absence of proteinuria. Hubertus et al., in 2015 reported a series of 22 patients with BWT

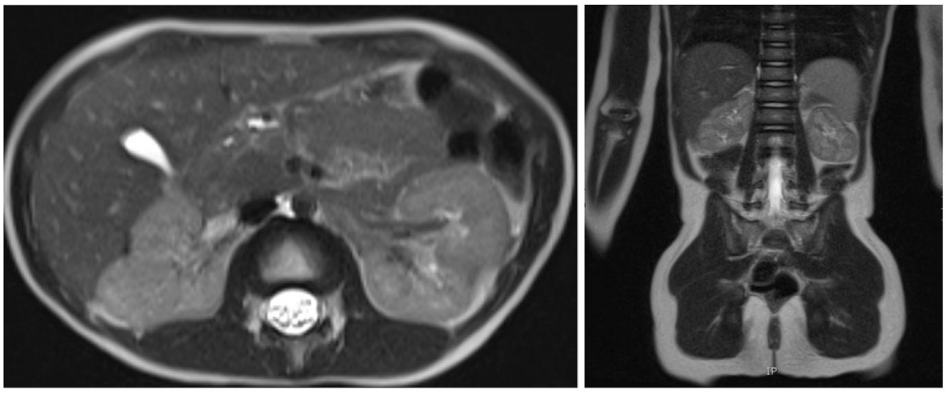

Fig. 3. Representative slices from MRI abdomen 2.25 years post-resection, 2.5 years post-presentation. Resolution of bilateral tumor burden with expected postsurgical changes of bilateral kidneys.

receiving either NSS or unilateral partial plus contralateral total nephrectomy. In this series overall survival and relapse rates were equal but hypertension was less common (66.7% vs 20%, $p = 0.043$) in patients receiving NSS.[4] Modern renal allotransplantation graft survival in a pediatric population at ten years ranges from 64 to 95%[5] and has the associated risks of long-term immunosuppression. This remains an undesirable course of treatment for the pediatric population, underlining the significance of preserving renal function in children with WT and BWT. In the adult population, preservation of 25–33% of a solitary renal unit has been shown to be sufficient to avoid dialysis or renal transplant, again motivating universal implementation of NSS for BWT in the pediatric population. In a 15 year series of BWT in Italy, 48 patients underwent bilateral NSS: at 5 years, survival was 80% and event free survival was 66.5%.[1] Here we present a case of extensive partial nephrectomies for BWT with completely event free, disease-free survival at three years.

Conclusion

NSS is a safe and oncologically therapeutic surgical approach in children presenting with BWT and can be considered even in patients with extensive tumor burden. We report medium-term outcomes for one such patient requiring resection of twelve oncologic lesions bilaterally with disease-free survival demonstrated by MRI at three years post-resection.

References

1. Kieran K, Davidoff AM. Nephron-sparing surgery for bilateral Wilms tumor. *Pediatr Surg Int Mar*. 2015;31(3):229–236. PMID:25633157 PMCID: PMC4336577.
2. Dome JS, Fernandez CV, Mullen EA, et al. On behalf of the COG renal tumors committee Children's Oncology Group's 2013 blueprint for research. *Renal Tumors Pediatr Blood Cancer*. 2013 Jun;60(6):994–1000.
3. Davidoff AM, Interiano RB, Wynn L, et al. Overall survival and renal function of patients with synchronous bilateral Wilms tumor undergoing surgery at a single institution. *Annals of surgery*. 2015;262.4:570–576. PMC. Web. 29 June 2017.
4. Hubertus J, Günther B, Becker K, et al. Development of hypertension is less frequent after bilateral nephron sparing surgery for bilateral Wilms tumor in a long-term survey. J Urol 193(1), 262–266.
5. Loiseau Y, Bacchetta J, Klich A, et al. Renal transplantation in children under 3 Years of age: experience from a singe-center study. *Pediatr Transplant*. 2018 Jan 17. https://doi.org/10.1111/petr.13116 (Epub ahead of print).