A Case of Prenatally Diagnosed Wilms Tumor Managed With Laparoscopic Nephrectomy

Amir Toussi, Candace F. Granberg, and Patricio C. Gargollo

We present a case of prenatally diagnosed Wilms tumor, with subsequent operative approach and management. The patient was referred to our institution with an abnormal prenatal renal ultrasound. Computed tomography scan after delivery showed a multifocal enhancing left renal mass. The patient underwent an uncomplicated left laparoscopic radical nephrectomy and retroperitoneal lymph node sampling using a novel hidden incision endoscopic surgery technique. Final pathology revealed favorable histology stage II Wilms tumor. The patient underwent adjuvant chemotherapy with vincristine and dactinomycin based on the EE4A regimen. We highlight the diagnostic pathway, perioperative management, surgical approach, and early postoperative follow-up in this case report. UROLOGY 113: 197–199, 2018. © 2017 Elsevier Inc.

Wilms tumor is the most common primary malignant solid renal tumor of the kidney in childhood. However, neonatal incidence is extremely low and prenatal detection is even more uncommon. In fact, according to the National Wilms' Tumor Study 1-3, only 0.12% of all Wilms tumors occurred in patients younger than 30 days old.[1] More recent series have identified that approximately 10% of prenatally detected solid renal tumors are Wilms tumors.[2,3] We present a case of prenatally diagnosed Wilms tumor and offer insight into a contemporary and minimally invasive surgical approach.

CASE PRESENTATION

The patient was a neonatal female referred to our institution when found to have an abnormal renal ultrasound on routine second-trimester prenatal ultrasound. The mass was seen on subsequent ultrasounds at 32 weeks and again at 37 weeks. Neonatal ultrasound confirmed the mass with the presence of Doppler flow, and a contrast-enhanced computed tomography scan performed on day 10 of life revealed a centrally located, multifocal, enhancing left renal mass (Fig. 1). The contralateral kidney, retroperitoneum, renal vein, and inferior vena cava were without any abnormalities. The remainder of the patient's staging workup, including a chest computed tomography, was negative.

The patient underwent a laparoscopic radical left nephrectomy with lymph node sampling on day 26 of life. She was placed in slight right lateral decubitus position using a small bump under her left flank. The surgical approach was initiated by placing 3 laparoscopic ports using hidden incision endoscopic surgery technique as described by Gargollo[4] (Fig. 2). The dissection was started by mobilizing the left colon medially. The ureter and the gonadal vein were identified and lifted up, and dissection was marched toward the hilum using hook electrocautery instrument. A single artery and vein were identified. The artery was triply ligated with Hem-O-Lock (Teleflex) clips and divided. Then the vein was clipped and divided in a similar fashion. Lymph node sampling of the hilar lymph nodes was performed using a combination of Hem-O-Lock (Teleflex) clips on the stay side and Harmonic Ace (Ethicon) shears to achieve lymphatic control. The specimen was then freed superiorly and laterally using the Harmonic Ace (Ethicon) shears. Once the kidney was free of all attachments, the ureter was clipped and divided close to the bladder. The specimen was placed in an Endo Catch (Covidien) bag. The 2 lower incisions (at the level of Pfannenstiel incision) were extended toward each other to create 1 larger incision, which accommodated the delivery of the specimen out of the abdomen. The total operative time was 98 minutes, whereas the pneumoperitoneum was used for 76 minutes.

There were no intraoperative complications or tumor spillage. Intraoperative frozen pathology was consistent with Wilms tumor; therefore, a central venous catheter was placed for administration of adjuvant chemotherapy.

The histopathologic examination revealed favorable histology stage II Wilms tumor due to extension into the renal sinus (Fig. 3). Surgical margins were negative and there was no lymph node involvement (0 of 1 sampled lymph nodes). The patient was dismissed on postoperative day 1 without any complications. She underwent adjuvant chemotherapy with vincristine and dactinomycin based on the

This is a Pediatric Urologic Oncology Working Group case submission that was solicited by Urology.

Financial Disclosure: *The authors declare that they have no relevant financial interests.*

From the Department of Urology, Mayo Clinic, Rochester, MN

Address correspondence to: Patricio C. Gargollo, M.D., Department of Urology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905. E-mail: gargollo.patricio@mayo.edu

Submitted: June 14, 2017, accepted (with revisions): October 3, 2017

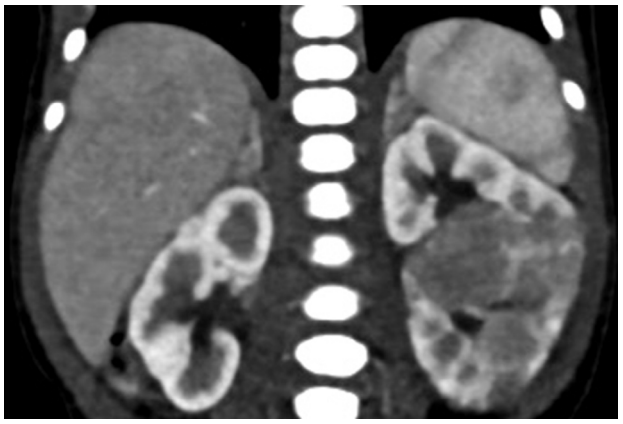

Figure 1. Computed tomography scan with intravenous contrast showing a multifocal centrally located enhancing renal mass confined to the left kidney.

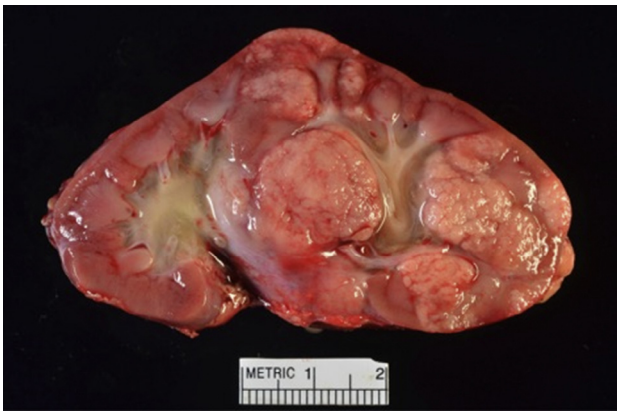

Figure 3. Gross examination revealing multifocal Wilms tumor with invasion into the renal sinus fat. (Color version available online.)

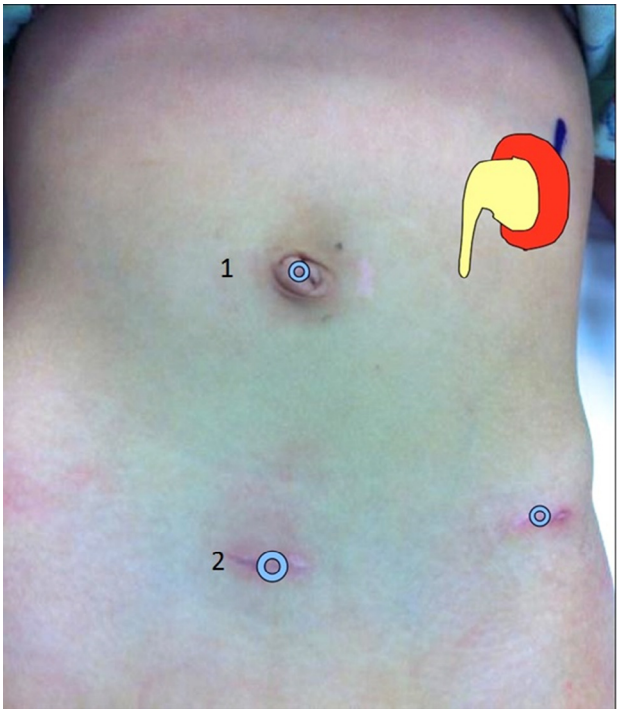

Figure 2. Hidden incision endoscopic surgery port placement for laparoscopic radical nephrectomy places the scars at the umbilicus and at the Pfannenstiel level. These images show the 3-month postoperative scar profile. Numbers 1 and 2 represent the left working port and the camera port respectively. (Color version available online.)

EE4A regimen.[5] She tolerated the full course of her chemotherapy and remained without evidence of disease at her 6-month follow-up.

DISCUSSION

Congenital mesoblastic nephroma is a far more common cause of solid renal mass than Wilms tumor in children under the age of 3 months. Furthermore, the reported incidence of prenatal detection is extremely low and the literature is limited to only case reports.[2,6-9] Besides ultrasonography, fetal magnetic resonance imaging has been helpful in diagnosis.[10] Currently, there is no role in fetal intervention for solid renal masses and definitive treatment is pursued after delivery.

In general, neonatal tumors tend to be more aggressive and Wilms tumors are oftentimes associated with complicating factors such as fetal hydrops, polyhydramnios, hypertension, or hypercalcemia.[2,10,11] Also, complications during delivery or in the immediate newborn period may result in further morbidity and mortality. Therefore, a multidisciplinary approach at tertiary care centers with the appropriate resources and expertise is necessary to best manage perinatal complications.

In the absence of complicating factors or after appropriate management of life-threatening abnormalities, these intrarenal solid tumors are best managed surgically by an experienced pediatric surgical team to achieve negative margins without tumor spillage. Surgical complications are reported to be as high as 25%, including tumor rupture, intraoperative bleeding, and death.[11] Outside of the critical perinatal period, these tumors tend to have excellent oncological outcomes. In a recent series, cancer-specific survival over a median follow-up period of 4 years reached 100%.[2,3,11] This finding highlights the importance of timely referral to appropriate care centers with available multispecialty pediatric teams upon detection of any prenatal renal mass.

In this case, the patient was referred to our institution in a timely fashion and was managed by a multidisciplinary team of pediatric subspecialists, resulting in a complication-free perioperative course. Of note, our patient's radical nephrectomy and lymph node sampling were performed via a minimally invasive approach, a novel technique known as hidden incision endoscopic surgery.[4] Although preserving oncological principles (negative surgical margins, lymph node sampling, no tumor spillage), this technique allows excellent cosmetic results. The incisions are

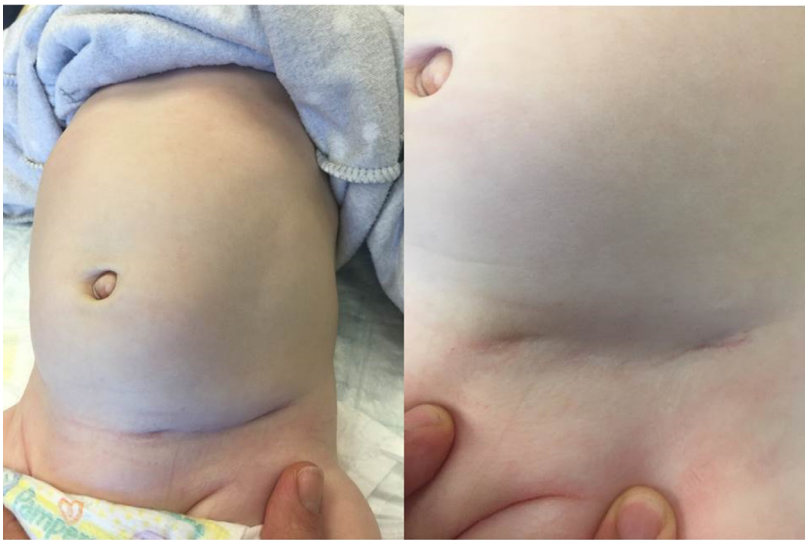

Figure 4. 3-Month postoperative scar profile using hidden incision endoscopic surgery technique. (Color version available online.)

hidden in the umbilicus and at the level of a Pfannenstiel incision, which render them nonvisible if the patient is wearing a bathing suit (Fig. 4).

Traditionally, the surgical approach in neonates has been via an open incision.[8,12] An open approach may be warranted if there is concern for tumor spillage, intraoperative complications, and inadequate resection. Other factors such as hemodynamic instability requiring minimal anesthetic exposure, the size of the primary tumor, and the patient's age may also contribute to the decision regarding an open surgical approach. Our decision was influenced by several facts including the availability of an experienced surgeon, small size of the tumor, an organ-confined disease, the absence of aberrant anatomy, and the absence of perinatal complications. In a large, multi-institutional series, minimally invasive nephrectomy for Wilms tumor was comparable with an open approach in regard to oncological control, perioperative complications, and overall survival.[13] It is important to note that the average age in that series was 40 months, although we can extrapolate the safety of the minimally invasive approach to the neonatal period in highly selected patients at experienced tertiary centers.

We acknowledge the limitations in performing an extensive lymph node dissection with the minimally invasive approach and only consider the approach in highly selected patients in order not to compromise oncological outcomes. There have been rare reports of air embolism from pneumoperitoneum in neonates during laparoscopic surgery. This is a rare complication; however, measures such as utilizing CO_2 gas insufflation and limiting insufflation times must be employed to mitigate the risk of air embolism.

CONCLUSION

Wilms tumor is a rare cause of a prenatally detected solid renal mass. Timely referral to a high-volume tertiary care center is important. The minimally invasive approach is a safe and viable surgical option in highly selected patients in the neonatal period.

References

1. Hrabovsky EE, Othersen HB Jr, deLorimier A, et al. Wilms' tumor in the neonate: a report from the National Wilms' Tumor Study. *J Pediatr Surg.* 1986;21:385.
2. Glick RD, Hicks MJ, Nuchtern JG, et al. Renal tumors in infants less than 6 months of age. *J Pediatr Surg.* 2004;39:522.
3. Raciborska A, Bilska K, Weclawek-Tompol J, et al. Solid cancers in the premature and the newborn: report of Three National Referral Centers. *Pediatr Neonatol.* 2016;57:295.
4. Gargollo PC. Hidden incision endoscopic surgery: description of technique, parental satisfaction and applications. *J Urol.* 2011;185:1425.
5. Green DM, Cotton CA, Malogolowkin M, et al. Treatment of Wilms tumor relapsing after initial treatment with vincristine and actinomycin D: a report from the National Wilms Tumor Study Group. *Pediatr Blood Cancer.* 2007;48:493.
6. Applegate KE, Ghei M, Perez-Atayde AR. Prenatal detection of a Wilms' tumor. *Pediatr Radiol.* 1999;29:65.
7. Ritchey ML, Azizkhan RG, Beckwith JB, et al. Neonatal Wilms tumor. *J Pediatr Surg.* 1995;30:856.
8. Sarin YK, Rahul SK, Sinha S, et al. Antenatally diagnosed Wilms' tumour. *J Neonatal Surg.* 2014;3:8.
9. Suresh I, Suresh S, Arumugam R, et al. Antenatal diagnosis of Wilms tumor. *J Ultrasound Med.* 1997;16:69.
10. Vadeyar S, Ramsay M, James D, et al. Prenatal diagnosis of congenital Wilms' tumor (nephroblastoma) presenting as fetal hydrops. *Ultrasound Obstet Gynecol.* 2000;16:80.
11. Leclair MD, El-Ghoneimi A, Audry G, et al. The outcome of prenatally diagnosed renal tumors. *J Urol.* 2005;173:186.
12. Jain V, Mohta A, Sengar M, et al. Is antenatal detection of Wilms' tumor a bad prognostic marker? *Indian J Med Paediatr Oncol.* 2011;32:214.
13. Warmann SW, Godzinski J, van Tinteren H, et al. Minimally invasive nephrectomy for Wilms tumors in children—data from SIOP 2001. *J Pediatr Surg.* 2014;49.