Pediatric Case Reports

Adult Wilms Tumor During Pregnancy: Case Report and Literature Review

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Adult Wilms tumor (WT) is a well-known, albeit rare entity and has historically been associated with worse overall clinical outcomes when compared to younger patients. Because WT is uncommon in adult patients, it is often misdiagnosed and treated off standardized pediatric protocols. WT associated with pregnancy is even more rare, and there is not a standardized approach to this small subset of patients. We present a case of an adult WT discovered and managed during the perinatal period and review prior published cases. UROLOGY 129: 200–205, 2019. © 2018 Elsevier Inc.

Adult Wilms tumor (WT) comprises approximately 3% of all WT diagnoses,[1] and was initially found to have worse overall survival rates.[2] However, outcomes of adults with WT continue to improve in the modern era when treated on similar protocols to children.[3,4] An even smaller subset of adult WT patients present during pregnancy. There are 11 reported cases in the English literature involving a WT diagnosis in the perinatal period. Of those, only 8 were diagnosed during the pregnancy, and only 4 received treatment while the patient was still pregnant. We present another case of WT diagnosed and treated during pregnancy.

CASE-PRESENTATION

A 23-year-old G1P0 female presented initially to her obstetrician at 14-week gestational age (GA) with right flank pain. Renal ultrasound revealed an 8 cm right, complex renal cyst. Magnetic resonance imaging (MRI) was used to better characterize this lesion, and due to a preliminary diagnosis of a complex cyst (benign), conservative management through delivery was recommended. Unfortunately, she developed worsening flank pain and by 18-week GA was referred to her local urologist. Triphasic computed tomography (CT) of the abdomen and pelvis was performed, which demonstrated interval growth of the lesion to 13 cm with associated hemorrhage. She was diagnosed with a bleeding angiomyolipoma, and underwent angioembolization of the mass. On follow-up MRI at 26-weeks GA, the renal mass had grown to 17 cm. Due to the concern that this represented a malignancy, she underwent staging chest CT and was found to have multiple pulmonary nodules (Fig. 1). Biopsy of the mass showed triphasic WT. At that time, she was referred to a tertiary care center with overall stage IV and local stage III WT.

A multidisciplinary conference was held to discuss the management of WT during the third trimester of pregnancy. Options reviewed included expectant management with restaging and treatment after delivery, prenatal radical nephrectomy (RN) followed by chemotherapy before or after delivery, and prenatal chemotherapy followed by postpartum RN. Ultimately the decision was made to proceed with scheduled cesarean-section (C/S) delivery at 28-week GA followed immediately by RN and lymph node dissection (LND) under the same anesthetic.

The patient underwent successful C/S delivery of a 980-g baby girl, Apgar 5 and 8 at 1 and 5 minutes respectively. The child was transferred immediately to the neonatal intensive care unit where she spent 42 days on nonventilator respiratory support. Due to the concern for possible transplacental tumor transmission, abdominal, and cranial ultrasounds were performed, both of which were negative for metastatic disease. The baby was discharged at adjusted term age. After delivery of the fetus, RN and LND were performed through a separate, hemichevron incision (Fig. 2). The patient tolerated the procedure well and was observed in the intensive care unit overnight. On postoperative day (POD) 1, she was transferred to the acute-care floor in stable condition. The remainder of her hospital course was unremarkable, and she initiated breast-feeding prior to discharge on POD 8.

Final pathology revealed a normal placenta and a 30 cm WT with capsular extension and focal anaplasia. Surgical margins were negative for tumor and 0/20 nodes were involved. Two weeks postoperatively she was started on Children's Oncology Group (COG) study AREN0321 protocol, which included 1 week of 10.5 Gy flank radiation and 12 Gy whole lung radiation followed by 30-week of combination regimen UH-1 chemotherapy (alternating high-dose cyclophosphamide [CP], vincristine [VCR], and doxorubicin [DOX] and low-dose CP, carboplatin, and

Conflicts of Interest: None.

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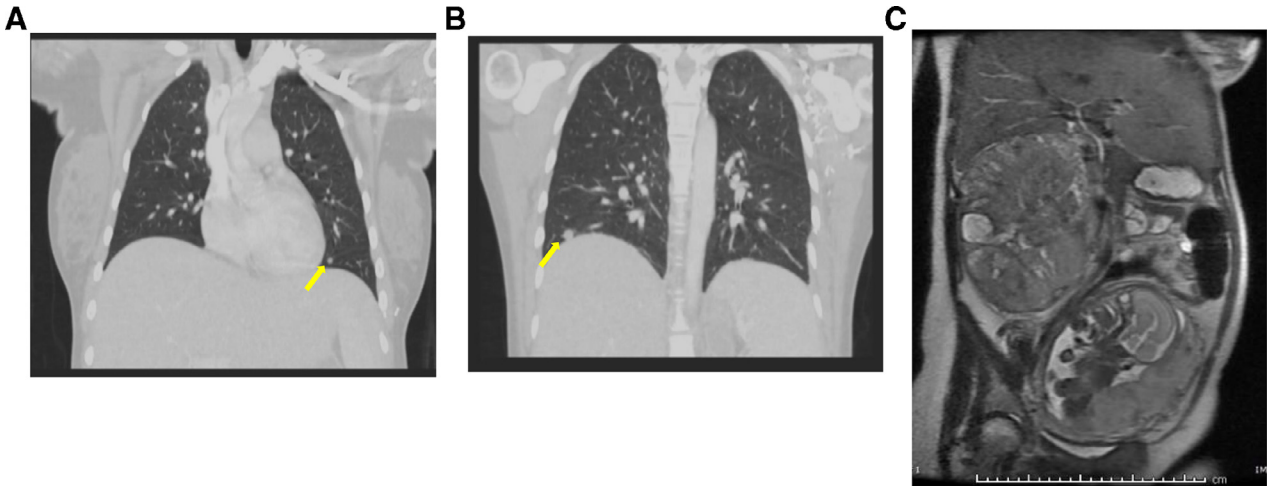

Figure 1. Computed tomography (CT) of the lungs revealing several lung nodules (yellow arrows) (A and B). Magnetic resonance imaging (MRI) of the abdomen and pelvis at 26-week gestational age demonstrating large right renal mass adjacent to fetus (C). (Color version available online.)

etoposide). She had a complete therapeutic response with resolution of the pulmonary metastases. Fourteen months postoperatively, she and her child remain disease free.

DISCUSSION

Outcomes for adults with WT are improving with early diagnosis and enrollment into treatment protocols based on the work done by COG. With the use of protocol driven multimodal therapy, recurrence rates, overall survival, and disease-specific survival rates have all drastically improved. More recent data have shown that 5-year overall survival for adults with favorable histology tumors was as high as 82.6%.[3] Like pediatric protocols, current recommendations for treatment of adult WT are based on tumor stage and histology. Some differences in adult WT protocols include introduction of radiation and doxorubicin in Stage II disease, and preoperative chemotherapy for patients diagnosed with biopsy.[5] This highlights the need for early recognition and proper staging in adult WT patients.

Adult WT is even more rare in the pregnant population. There are only 8 reported cases in the English literature where the diagnosis was made prenatally, and only 4 of those patients received any treatment prior to delivery of the fetus. All prenatal diagnoses were made during the second or third trimester with all stages seen. Reported follow-up ranged from 4 weeks to 11 years, with 2 deaths. Unfortunately, only 2 case reports include any long-term follow-up regarding the child (Table 1).

Malignancies diagnosed during pregnancy are rare, with an estimated incidence of 1-2 cases per 1000 pregnancies.[6]

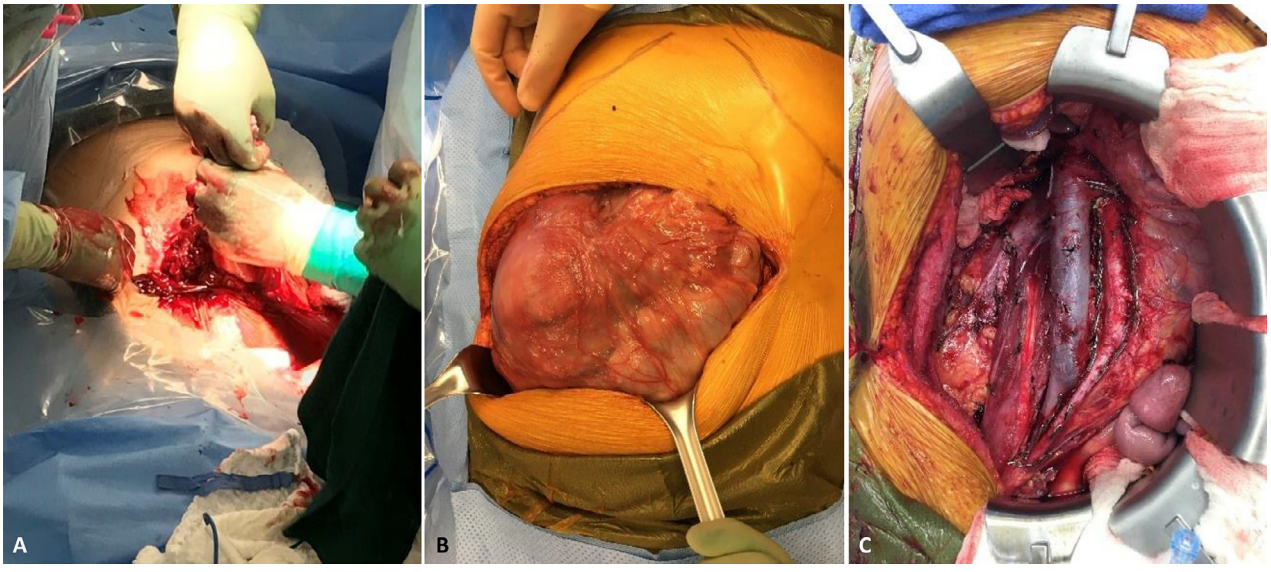

Figure 2. Cesarean section through a Pfannenstiel incision (A). Large right renal tumor extruding through hemi-chevron incision (B). Retroperitoneum post-tumor resection and lymph node dissection (C). (Color version available online.)

Table 1. English literature case reports of Wilms tumor associated with pregnancy.

Title	First Author	Year	Age (yrs)	Timing of Diagnosis	Presenting Symptom	COG Stage*	Treatment	Patient Follow-up	Baby Follow-up
Wilms's tumor in the adult; review of literature and report of three additional cases	Eserky, G	1947	24	7 mo Postpartum	Left flank pain ("enlarged spleen" noted during pregnancy)	*II*	1. Postpartum RN 2. Postpartum RTx: "15 deep x-ray treatments"	3 yrs; NED	None
Wilm's tumor in an adult: report of a 10-year cure†	Livermore, G	1953	37	Prenatal ("loin mass" noted 3yrs prior during pregnancy)	Flank pain	*III*	1. Postpartum tumor biopsy 2. Postpartum RTx: tumor (4100 rads) 3. Postpartum RN	11 yrs; NED	None
Wilms' tumor complicating pregnancy: report of a case†,‡	Davis, L	1987	19	32 wks GA	Gross hematuria	*I or II*	1. Prenatal RN 2. Postpartum CTx: DACT and VCR	15 mo; metastatic disease	None
Adult Wilms' tumor: Prognostic and management considerations†	Bozeman, G	1995	21	32 wks GA	Flank pain	IV	1. Postpartum RN 2. Postpartum CTx: 3 cycles IFO, CBDCA, EPEG 3. Postpartum RTx: whole abdomen (3000 rads), lungs (1200 rads) 4. Postpartum lung wedge resection 5. Postpartum alternative CTx: VCR, DOX, and DACT	40 mo; NED	None
An unusual tumor in a post-partum woman	Singh, N	1999	25	6 mo postpartum	Abdominal swelling	*IV*	1. Postpartum tumor biopsy 2. Refused treatment	4 wks, Deceased	None
Second pregnancy-associated Wilms tumor 16 years after the first one	Wynn, T	2003	19	1. Immediately postpartum	1. Abdominal mass	1. *V (Bilateral)*	1. First Presentation: a. Postpartum biopsy b. Postpartum CTx: VCR, DACT, DOX c. Postpartum RN and contralateral renal wedge biopsy	1. 5 yrs; NED, lost to follow-up	1. None
				2. 16 yrs later, immediately postpartum	2. Abdominal mass	2. *III*	2. Second Presentation: a. Postpartum tumor biopsy	2. 6 mo, deceased	2. None

Continued

Table 1. Continued

Title	First Author	Year	Age (yrs)	Timing of Diagnosis	Presenting Symptom	COG Stage*	Treatment	Patient Follow-up	Baby Follow-up
							b. Postpartum CTx: VCR, DACT, DOX c. Postpartum RN d. Postpartum RTx: Intra-operative to tumor bed (1500 cGy)		
Spinal cord compression and lung metastasis of Wilms' tumor in a pregnant adolescent[†,‡]	Carapcioglu, F	2004	19	1. 8 mo prior to pregnancy 2. 25 wks GA	1. Palpable mass 2. RLE weakness	1. *II* 2. *IV*	1. First presentation: a. RN 2. Second presentation: a. Prenatal CTx: VCR and DACT b. C/S delivery (28-week) c. Postpartum RTx: Tumor bed and spine (3000 cGy) d. Postpartum CTx: 5-cycles IFO, CBDCA, EPEG alternated with VCR, DACT, CP	1. Lost to follow-up 2. 11 mo; NED	1. N/A 2. 10 mo; NED
Wilm's tumor during pregnancy: report of laparoscopic removal and review of literature[†]	Rehman, J	2008	36	3rd Trimester	Gross hematuria	II	1. Postpartum laparoscopic RN 2. Postpartum CTx: 18wks VCR and DACT	18 wks (no mention of disease status)	None
Multimodal tumor therapy in a 31-year-old pregnant woman with Wilms tumor[†,‡]	Maurer, T	2009	31	18 wks GA	Hyperemesis gravidarum	II	1. Prenatal RN (19 wks GA) 2. Prenatal CTx (22 wks GA): VCR and DACT 3. C/S delivery (33 wks GA) 4. Postpartum CTx: 6-cycles VCR, DACT, DOX	4 yrs; NED	4 yrs; NED
Adult Wilms tumor during gestational period[†,‡]	Rodrigues, F	2009	17	20 wks GA	Gross hematuria	I	1. Prenatal RN 2. Refused CTx	2 yrs; NED	None

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Table 1. Continued

Title	First Author	Year	Age (yrs)	Timing of Diagnosis	Presenting Symptom	COG Stage*	Treatment	Patient Follow-up	Baby Follow-up
Wilms tumor: an uncommon entity in the adult patient[†]	Mahmoud, F	2016	28	Prenatal (mass originally seen 2yrs prior on ultrasound during pregnancy)	Incidental	*III*	1. Postpartum laparoscopic RN 2. Postpartum CTx: DACT, VCR, and DOX	2 yrs; NED	None

CBDCA, carboplatin; C/S, cesarean section; CP, cyclophosphamide; CTx, chemotherapy; DACT, actinomycin-D; DOX, doxorubicin; EPEG, etoposide; GA, gestational age; IFO, ifosfamide; Mo, months; NED, no evidence of disease; RN, radical nephrectomy; RTx, radiotherapy; VCR, vincristine; Wks, weeks; Yrs, years.
† Prenatal diagnosis.
‡ Patient received therapy in the prenatal period.
* Italics denote Stage was not explicitly noted in original text and was interpreted based on information in case report.

Renal tumors compose a small minority of these malignancies, and no specific approach to management is recommended. Initial staging of a pregnant patient with a suspected renal malignancy ideally should be done with nonionizing radiation, most commonly with ultrasound and/or MRI. If a malignancy is diagnosed, considerations as to the approach of local and systemic therapy should be made regarding how the treatment will affect both the mother and fetus.[7]

Excluding appendectomy, pregnant and nonpregnant women undergoing nonobstetric surgery have similar morbidity and mortality rates.[8] A meta-analysis of the effects of surgery during pregnancy on maternal and fetal outcomes reported exceedingly rare maternal death rates (0.006%), miscarriage (5.8%), and premature labor (3.5%).[9]

Currently, chemotherapy regimens delivered after the first trimester are considered relatively safe, but multidisciplinary discussions and appropriate patient counseling should be held prior to initiation of treatment.[10] Aviles and Neri reported on 84 children born to mothers with a hematologic malignancy diagnosed and treated during pregnancy, 38 during the first trimester. Several chemotherapeutics used in WT protocols were included (CP, VCR, DOX). At a median follow-up of 18.7 years, these children were noted to have normal educational performance, no congenital or neurologic abnormalities, and no reported malignancies.[11] However, because the NCCN recommends both VCR and actinomycin-D for the treatment of gestational trophoblastic tumors,[12] we chose to avoid these agents prepartum in the present patient over concerns for risk to the placenta.

Reported follow-up for pregnancy associated WT patients is sparse. Current follow-up recommendations for adult WT patients include monitoring for treatment toxicity (liver, lung, and cardiac) and routine chest and abdominal imaging surveillance every 3 months for at least 2 years.[5] There is even less reported follow-up of the children born to women pregnant during the initial WT diagnosis. Although there are no reports of maternal-fetal metastasis of WT, transplacental metastases have been reported with leukemia, melanoma, and lung cancer.[13] Since no official screening recommendations for infants born to mothers with cancer exist, we chose surveillance with whole-body MRI and chest CT at 6 months of age, followed by a symptom-based approach.

CONCLUSION

Adult WT in pregnancy is exceedingly rare with few reported cases. Outcomes of patients presenting as adults with WT are improving with standardized treatment regimens based on pediatric protocols. In the modern era, surgery and even chemotherapy are options in properly chosen and counseled pregnant patients with a WT.

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