Case - Adult Wilms tumor in pregnancy: Incidental first-trimester discovery, antepartum radical nephrectomy, and third-trimester adjuvant chemotherapy

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INTRODUCTION

Wilms tumour (nephroblastoma) is the most common pediatric renal malignancy with a peak incidence at 3-4 years of age. Adult Wilms tumour is rare, accounting for fewer than 1% of adult renal malignancies and is historically associated with more advanced disease at diagnosis, with inferior outcomes compared to the pediatric population.[1,2] The co-occurrence of adult Wilms tumour with pregnancy is exceptionally rare. Walker et al. previously identified 11 prior English-language case reports diagnosed in the perinatal period in 2018; four additional cases have been published since then.[1,3–6]

KEY MESSAGES

- Adult Wilms tumour in pregnancy is exceptionally rare; incidental discovery at 11 weeks' gestation on routine dating ultrasound represents the earliest documented gestational diagnosis in the published literature
- Open radical nephrectomy at 20 weeks' gestation was feasible and uncomplicated; adjuvant chemotherapy administered from 27 weeks' gestation throughout the third trimester resulted in a healthy term delivery at 38+6 weeks (Apgar 9/9) without neonatal complications
- Molecular profiling identifies a pathogenic CTNNB1 p.S33F mutation and adverse 17p13.1 loss of heterozygosity in a histologically favourable tumour, demonstrating genomic risk stratification can prompt treatment escalation beyond histology alone in adult Wilms tumour

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Herein, we report a case of adult Wilms tumour discovered incidentally on first-trimester dating ultrasound, managed with antepartum radical nephrectomy and third-trimester adjuvant chemotherapy, with comprehensive molecular profiling, resulting in a healthy term delivery and no evidence of disease at two-years of follow-up.

CASE REPORT

A 31-year-old primigravid woman (G1P0) was found to have an incidental large heterogeneous left upper quadrant mass (8.1×6.4×19.9cm) arising from the left kidney at 11 weeks' gestation during a routine first-trimester dating ultrasound. She was asymptomatic with no hematuria, flank pain or constitutional symptoms. MRI of the abdomen (Figure 1) demonstrated a 16cm multilobulated heterogeneous left renal mass with associated hydronephrosis and a borderline left periaortic lymph node measuring 1.3×0.9 cm. Staging CT of the chest and brain was negative for distant metastatic disease.

At 20 weeks' gestation, the patient underwent open adrenal-sparing transperitoneal left radical nephrectomy with paraaortic lymph node dissection. The obstetric service was notified preoperatively and fetal surveillance was conducted postoperatively without complication. Intraoperatively, a large multilobulated renal mass was identified without gross invasion of adjacent structures or renal vein tumour thrombus. Small paraaortic lymphadenopathy was noted intraoperatively, prompting a formal lymph node dissection. Estimated blood loss was 100 mL with no perioperative complications.

Surgical pathology of the nephrectomy specimen (Table 1; Figures 2-4) revealed a large renal tumour measuring 16.0×13.5×13.5 cm and weighing 1601.2 grams. It demonstrated a biphasic, epithelial-predominant Wilms tumour showing approximately 50% spontaneous necrosis. Histology of viable tumour showed predominantly tubulopapillary architecture, poorly differentiated areas with focal squamous differentiation and hyalinized stroma with scattered dystrophic calcifications. No unequivocal cytologic anaplasia was identified, indicating Wilms tumour favourable histology. Pathologic staging features included focal capsular penetration, renal sinus adipose tissue invasion, a suspicious positive hilar surgical margin and focal small vessel lymphovascular invasion. Eight paraaortic lymph nodes were tumour-free (0/8); several nodes contained ectopic decidual tissue consistent with the gravid state. Nephrogenic rests were absent in the residual non-neoplastic kidney. By Children's Oncology Group (COG) criteria, the tumour was classified as definite Stage II based on the renal sinus involvement and considered Stage III based on the suspected positive hilar margin.

Immunohistochemistry demonstrated diffuse nuclear PAX8 and pan-cytokeratin positivity with WT1 negativity – a pattern present in a recognized subset of Wilms tumours.[7] p53 expression was wild-type by immunohistochemistry. Foci of decidual tissue in paraaortic lymph nodes were positive for ER and PR and negative for pan-cytokeratin. A 31-gene hotspot next-generation sequencing panel identified a pathogenic CTNNB1 p.Ser33Phe (c.98C>T) mutation at 39% allelic fraction, reflecting Wnt/β-catenin pathway activation.[7] Chromosomal microarray (Affymetrix OncoScan™ CNV; Nationwide Children's Hospital) identified 17p13.1 loss of heterozygosity (LOH), an adverse-risk genomic feature associated with high recurrence risk

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linked to TP53 pathway deregulation, in the setting of histologically favourable disease.[8] Chromosomal regions 1p, 1q, 11p15 and 16q were unaffected.

Following multidisciplinary discussion with urology, pathology, obstetrics and medical oncology, and in accordance with the patient's informed wishes, adjuvant DD-4A chemotherapy (vincristine, actinomycin-D, doxorubicin) was initiated at approximately 27 weeks' gestation. Upon identification of 17p13.1 LOH, the regimen was escalated to COG Regimen M (vincristine, etoposide, cyclophosphamide); one cycle was completed by approximately 35 weeks' gestation. Fetal growth surveillance by ultrasound remained appropriate throughout the third trimester. Labour was subsequently induced at 38+6 weeks for gestational hypertension (managed with nifedipine) and for oncologic indications. The patient delivered a healthy female infant via spontaneous vaginal vertex delivery (birthweight 3.11 kg; Apgar 9 and 9 at one and five minutes, respectively). No neonatal resuscitation was required. Postpartum hemorrhage (estimated blood loss of 600mL) was managed with oxytocin without further intervention. Following delivery, the patient declined the remaining six months of adjuvant chemotherapy and adjuvant radiotherapy after thorough counselling regarding long-term toxicity risks and the absence of robust adult-specific outcome data. In latest follow-up in March of 2026, CT of the chest, abdomen and pelvis continues to demonstrate no evidence of recurrence or metastatic disease with plans for continued surveillance until 2028. The patient is currently disease-free with normal renal function and with a healthy infant who is developing normally.

DISCUSSION

This case contributes several clinically important observations to the sparse literature on adult Wilms tumour in pregnancy. First, incidental detection at 11 weeks' gestation on routine first-trimester dating ultrasound represents the earliest documented gestational diagnosis in the published literature. All prior prenatal diagnosis, including those reviewed by Walker et al. and the four subsequently published cases, were diagnosed in the second or third trimester, typically precipitated by gross hematuria, an enlarging abdominal mass or flank pain.[1,3–6] Routine first-trimester obstetric ultrasound represents a previously unrecognized diagnostic opportunity for this rare entity.

Second, this case demonstrates the feasibility of open radical nephrectomy at 20 weeks' gestation. The midline transperitoneal approach provided adequate exposure for a 1601.2-gram specimen with an estimated blood loss of 100 mL and no perioperative complications. The second trimester is the preferred window for oncologic surgery in pregnancy as organogenesis is complete and uterine size has not yet significantly compromised surgical access.[9] Among prior published cases, the majority underwent surgery in the postpartum period; Maurer et al. and Rodrigues et al. each performed prenatal radical nephrectomy in the second trimester with similar safety.[10,11]

Third, adjuvant chemotherapy was administered from approximately 27 weeks' gestation through 35 weeks, encompassing cytotoxic agents across two sequential regimens, without fetal compromise. Prenatal chemotherapy for pregnancy-associated adult Wilms tumour has been documented. Carapcioglu et al. initiated emergency vincristine and actinomycin-D at 25 weeks

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for cauda equina compression, achieving delivery at 28 weeks without fetal toxicity; Maurer et al. administered planned prenatal vincristine and actinomycin-D from 22 weeks with caesarean section at 33 weeks and a healthy male infant; Brown et al., Graham et al. and Sepenu et al. each documented prenatal cytotoxic therapy without fetal compromise.[1,3,4,6,10] The available evidence supports feasibility of post-organogenesis chemotherapy with appropriate multidisciplinary oversight, though risks of intrauterine growth restriction, preterm labour and neonatal myelosuppression remain.[9]

Fourth, this is the first case of adult Wilms tumour in pregnancy to undergo antepartum molecular profiling with results directly driving treatment escalation. The identified CTNNB1 p.S33F mutation reflects Wnt/β-catenin pathway activation, a subtype enriched in epithelial-predominant Wilms tumours.[7] The 17p13.1 LOH is particularly significant in a histologically favourable tumour. TP53 pathway alterations at 17p are strongly linked to anaplasia but have also been identified in non-anaplastic fatal tumours, conferring adverse prognostic risk independent of histologic grade.[8] COG has established chromosomal LOH, particularly at 1p and 16q, as an independent adverse prognostic factor in favourable-histology Wilms tumour. 17p LOH carries analogous prognostic weight supported by emerging molecular evidence.[8,12,13]

Management of adult Wilms tumour extrapolates from COG pediatric protocols. Kalapurakal et al. reported 82% overall survival among 45 adults with favourable-histology Wilms tumour treated in the modern era (1979–2001), with stage-specific survival of 100%, 92%, 70% and 73% for Stages I–IV respectively. Notably, 13% of patients enrolled on NWTS 4–5 died of dactinomycin-induced hepatic venoocclusive disease – a toxicity that may be underrecognized outside pediatric oncology.[2] The patient's decision to decline remaining adjuvant therapy reflects the genuine uncertainty in counseling adults where long-term toxicity data are absent. She remains without evidence of disease at two years but continued surveillance is warranted given the elevated recurrence risk conferred by 17p13.1 LOH.[8]

DRAFT

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FIGURES AND TABLES

Figure 1. (A) Coronal and (B) axial T2-weighted MRI demonstrating a 16cm heterogeneous multilobulated left renal mass at 18 weeks' gestation. The gravid uterus with an intrauterine fetus is visible inferiorly (A).

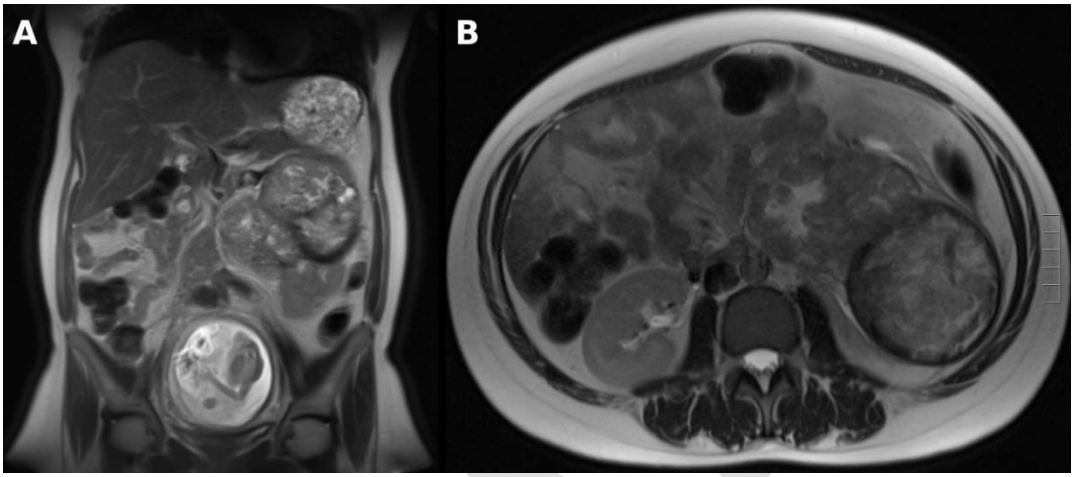
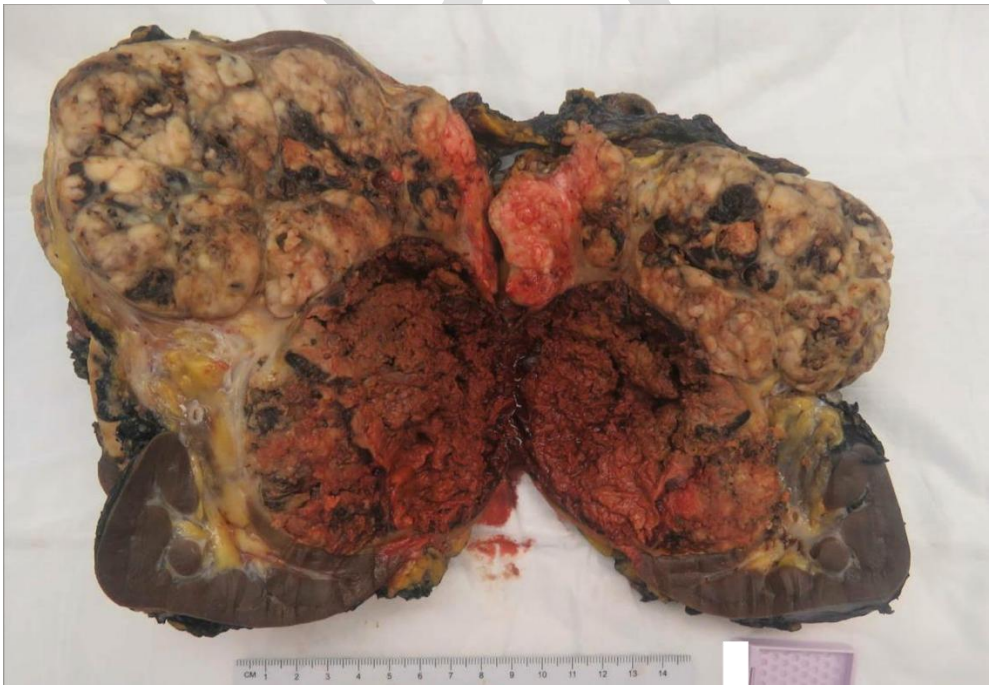


Figure 2. Gross pathology of the left radical nephrectomy specimen (1601.2g; 16.0×13.5×13.5 cm). Bivalved specimen demonstrates a multilobulated mass with heterogeneous tan-white solid nodules, approximately 50% spontaneous necrosis (dark areas) and focal hemorrhage. Residual non-neoplastic kidney is identifiable at the inferior pole.



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Figure 3. H&E stain; x200. Histology of the renal tumour shows a predominantly epithelial biphasic Wilms tumour with tubulopapillary architecture merging with foci of solid sheets of poorly differentiated epithelial malignancy and occasional foci of squamous differentiation. Stromal elements had focal calcifications.

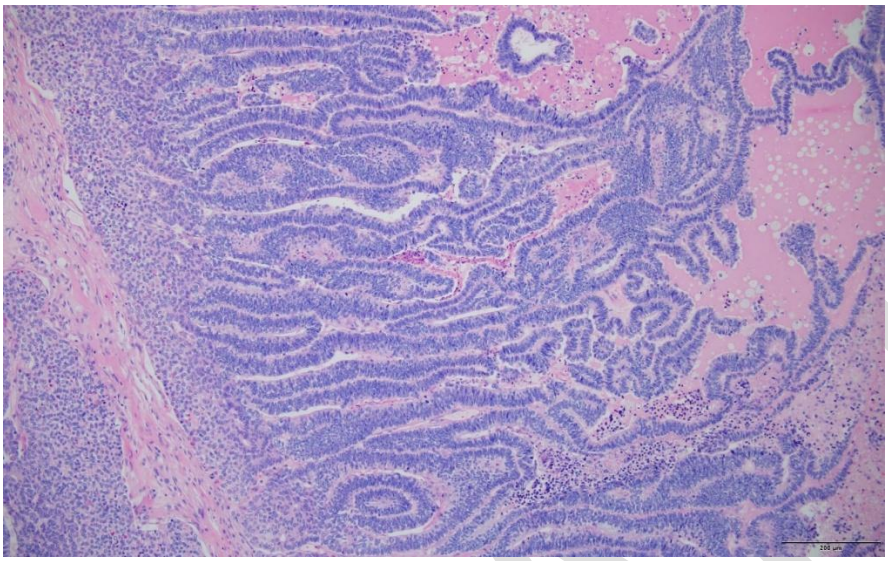
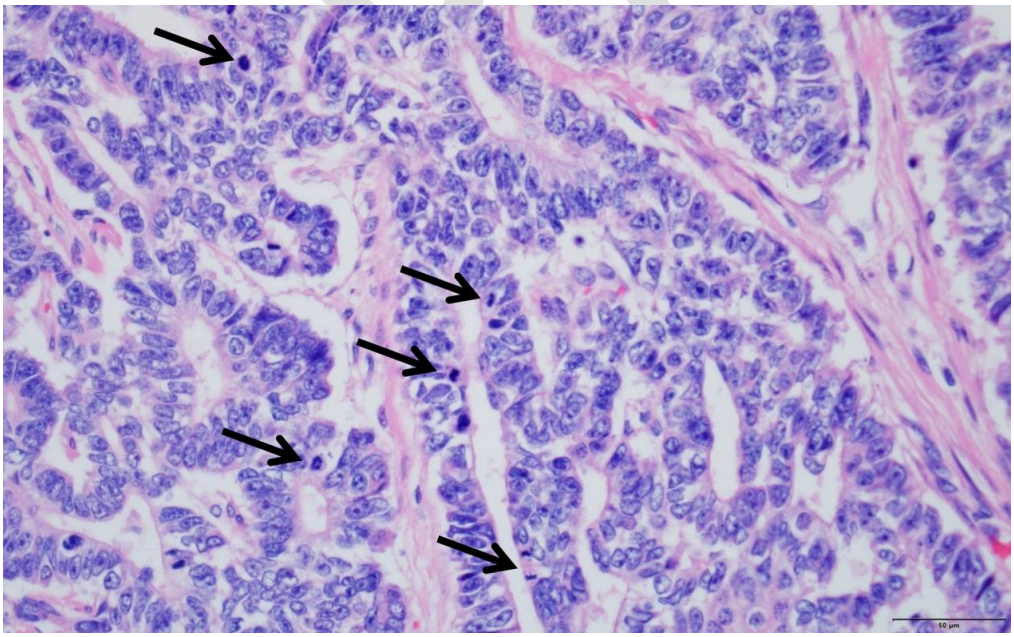


Figure 4. H&E stain; x400. The epithelial component shows Wilms tumour favourable histology; nuclei have variably prominent nucleoli and there are abundant normal appearing mitotic figures (arrows). The tumour is negative for cytologic anaplasia (marked nuclear enlargement, hyperchromasia and atypical mitotic figures).



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Table 1. Pathologic and molecular summary	
Parameter	**Findings**
Procedure	Open adrenal-sparing transperitoneal left radical nephrectomy + paraaortic lymph node dissection
Specimen weight	1601.2 g
Tumour size	16.0×13.5×13.5 cm
Histologic type	Wilms tumour, biphasic, epithelial-predominant; favourable histology (no anaplasia)
Architecture	Tubulopapillary with focal solid sheets; focal squamous differentiation with single-cell keratinization; hyalinized stroma with dystrophic calcifications; ~50% spontaneous necrosis
Blastemal component	Not identified
Extent of invasion	Capsular penetration; probable renal sinus adipose invasion; small vessel lymphovascular invasion; no large vein invasion; no pelvicalyceal mucosal involvement
Surgical margins	Suspicious positive hilar margin; perinephric soft tissue and vascular margins negative
Lymph nodes	0/8 paraaortic nodes positive; ectopic decidual tissue in several nodes (PR+, ER+, CD10+, CK-)
Nephrogenic rests	Absent
COG Stage	Stage III (positive surgical margin)
IHC – Positive	Pan-CK (AE1/AE3) diffuse; PAX8 diffuse; BCL1; BCL2; CK7 focal; P40 focal; CD117 focal; CAIX focal; CD56/CD57 focal; INI1 retained
IHC – Negative	WT1; p53 (wild-type expression pattern); AMACR; desmin; CD99; MART1; synaptophysin; SALL4; BRAF
NGS (Q31 hotspot panel)	CTNNB1 p.Ser33Phe (c.98C>T, exon 3); allelic fraction 39%; AMP/ASCO/CAP Tier 2C Pathogenic; TP53 and BRAF wild-type
Chromosomal microarray (OncoScan™ CNV)	17p13.1 LOH — POSITIVE (adverse-risk feature); 1p, 1q, 11p15, 16q — all negative

COG: Children's Oncology Group; IHC: immunohistochemistry NGS: next-generation sequencing; LOH: loss of heterozygosity.

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