WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
Coden USA: WJPRAP Impact Factor 8.63
Volume 15, Issue 10, 1188-1195. Case Study ISSN 2277–7105

WILM'S TUMOR IN 8-MONTH INFANT: A RARE CASE MANAGED WITH SIOP PROTOCOL AND NEPHRECTOMY

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Article Received on 14 April 2026,
Article Revised on 04 May 2026,
Article Published on 16 May 2026,

https://doi.org/10.5281/zenodo.20205949

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How to cite this Article: Dr. Bommagani Pavan Kumar*[1], Dr. Gaja Sai Chaitanya[1], Shaik Muneef[2], Dr. Shaik Safi Almas[3]. (2026). Wilm's Tumor In 8-Month Infant: A Rare Case Managed With Siop Protocol And Nephrectomy. World Journal of Pharmaceutical Research, 15(10), 1188–1195.

ABSTRACT

Wilm's tumor (nephroblastoma) is the most common pediatric renal malignancy, typically occurring in children under 5 years of age, with peak incidence between 3–5 years. We report a rare case of an 8-month-old infant who presented with complaints of swelling over the left lumbar region for 1 week, which was gradually increasing and associated with reduced urine output and multiple episodes of vomiting. Imaging with ultrasonography (USG) abdomen and contrast-enhanced computed tomography (CECT) confirmed the diagnosis. The patient was managed using a multidisciplinary approach following the SIOP protocol, including preoperative chemotherapy, followed by nephrectomy and postoperative chemotherapy. The postoperative period was uneventful, and the patient is currently under follow-up with stable clinical condition. This case highlights the importance of early diagnosis in atypical age groups and emphasizes that Wilm's tumor, although uncommon in infants, should be considered in the differential diagnosis of abdominal masses. Early intervention with a structured multidisciplinary approach can significantly improve prognosis.

KEYWORDS: Wilms tumor, pediatric renal cancer, SIOP Protocol, Nephrectomy.

INTRODUCTION

Wilm's tumor is the fourth most common pediatric cancer.[1] It affects approximately 1 in 10,000 children worldwide.[2] Wilm's' tumors occur sporadically, with a family history significant for the condition seen in just 1%-2% of patients.[3] Wilm's' tumor usually arises from embryonal kidney tissue, displaying several histological features that reflect its developmental origin. The tumor is often unilateral, but bilateral cases may occur in about 6% of patients.[4] Wilm's tumor most commonly manifests as the presence of abdominal pain, Systemic symptoms consist of fever, weight loss and fatigue, and less frequently also hematuria and hypertension.[5] Children with Wilm's tumor who received the SIOP-2001 protocol with preoperative chemotherapy the overall survival (OS) were seen to be 87%.[6] The overall multidisciplinary treatment survival rate is 90%, whereas it was only 30% with surgery alone and only 47% with the combination radiotherapy-surgery[7,8,9] The management of Wilm's tumor is centered on surgical resection. However, treatment strategies differ internationally: the Children's Oncology Group (COG), originating from the National Wilm's Tumor Study (NWTS) group in North America, recommends initial surgery followed by chemotherapy, while the International Society of Pediatric Oncology (SIOP) favors preoperative chemotherapy prior to surgical intervention.[10]

CASE REPORT

Basic information of patient:

8 MONTHS / FEMALE, 8.6 kgs.

History of present illness

Chief complaints

An 8-month-old baby girl presented with complaints of swelling over left lumbar region Since 1 week associated with reduced urine output and multiple episodes of vomiting upon oral intake of liquids soft diet and swelling gradually increasing in size.

History of past illness

Nil

Immunization Status

Up to date

Developmental History

Normal

Clinical Status at Admission

Stable

Family history

Nil

General examination

- The patient was conscious, oriented, and cooperative
- Well-nourished, moderately built
- No pallor, icterus, cyanosis, clubbing, pedal edema, or lymphadenopathy
- No signs of dehydration or cachexia.

Systemic Examination

Cardiovascular system

- Heart sounds are heard clearly and are normal with no murmurs.
- The pulse in both wrists (radial arteries) is felt at the same time indicate normal blood flow to both arms.
- The wrist pulse and groin pulse (femoral artery) occurs at the same time indicates normal blood flow

Respiratory system:

- Normal bilateral Air Entry with Normal vesicular breath sounds.

Central nervous system:

- The neurological findings are normal with no focal neurological deficits.

Abdominal examination:

- Soft, non-tender and bowl sounds are present from heart to the lower body.

LABORATOTY INVESTIGATIONS

USG ABDOMEN

USG Abdomen shows Left kidney enlarged in size of 100x97mm and a large heterogeneous soft tissue mass lesion arising from the left kidney which is well defined solid tumor mass with no involvement of middle and lower poles which suggests nephroblastoma (Wilm's tumor)

ALPHA FETO PROTEIN: 21.20 G/ML (Mild elevated)

CECT ABDOMEN

Large Well Defined Soft Tissue Density Lesion Noted at Left Upper and Mid Abdomen (35-40 HU) Arising from Left Kidney with Significant Compression of Renal Parenchyma Along

the Posteromedial Aspect of The Lesion. No Areas of Calcifications or Fat Density Are Identified with in the Lesions.

POST CONTRAST SCAN

Post-contrast imaging demonstrates a large, well-defined left renal mass lesion showing mild heterogeneous enhancement (55–60 HU). The lesion measures approximately **8.8 × 9.0 × 11.2 cm (transverse × anteroposterior × craniocaudal)** and extends across the midline. The mass effect is evident with medial displacement of the bowel loops and anterior branches of the aorta. The body and tail of the pancreas, along with the spleen, are displaced superiorly. The pelvicalyceal system of the left kidney is not dilated. The left ureter appears normal in caliber with prompt contrast excretion on delayed phase imaging. The left renal vessels are normal in course and caliber. No evidence of calcification, vascular invasion, or invasion into adjacent organs is identified. There is no significant lymphadenopathy.

WHOLE BODY PET CT

ABDOMEN

A large FDG-avid, heterogeneous abdominal mass is noted arising from the lateral cortex of the left kidney, measuring approximately **8.6 × 8.5 × 10.2 cm**, with a maximum standardized uptake value (**SUVmax**) of 5.4. No evidence of calcifications is identified within the lesion. The splenic vessels and left renal vessels are seen splayed over the mass. Anterolaterally, the lesion abuts the abdominal wall. Superiorly, it displaces the stomach as well as the body and tail of the pancreas medially. Superolateral, the lesion is closely abutting the inferior aspect of the spleen. Medially, the mass crosses the midline, causing displacement of the abdominal vessels, left adrenal gland, and head of the pancreas toward the right side. Adjacent bowel loops are displaced by the lesion. Inferiorly, the mass extends up to the iliac fossa.

IMPRESSION

FDG – PET CT scan findings show a large hyper metabolic heterogenic abdominal mass is noted arising from the lateral cortex left kidney crossing the midline with extent as described above, likely Wilm’s tumor. No other metabolically active lesion elsewhere in the regions of the body surveyed.

2D ECHO

Echocardiography showed normal cardiac anatomy and function, with situs Solitus, levocardia, intact Septae, no congenital defects, and no evidence of Coarctation of the aorta.

DIAGNOSIS

WILMS TUMOR (LOCALIZED) TREATMENT NEOADJUVANT CHEMOTHERAY: WEEK 1-3 CHEMOTHERAPY WITH VINCRISTINE+DACTINOMYCIN

Vincristine is given weekly, and Actinomycin D is administered intermittently, often alternating with vincristine-only weeks in neoadjuvant chemotherapy.

SURGERY: LEFT NEPHRECTOMY

AFTER NEOADJUVANT CHEMOTHERAPY

HISTOPATHOLOGY REPORT

Gross examination revealed a unifocal tumor measuring **13.5 cm** and weighing **975 grams**. Microscopy confirmed **Wilms tumor** with extensive rhabdomyoblastic differentiation. Post-chemotherapy changes were noted. The tumor was confined to the kidney with:

- Renal sinus: free
- Renal capsule: free
- Renal vein: free
- Renal pelvis: free
- No tumor rupture
- No lymph vascular invasion

The tumor was classified as **intermediate risk**, with viable tumor present and unfavorable histology components comprising less than 66% of viable tumor.

TREATMENT PROTOCOL

PLANNED FOR NEOADJUVANT WEEKLY CHEMOTHERAPY WITH VINCRISTINE+DACTINOMYCIN X 4 CYCLES.

Baby tolerated the chemotherapy well with no adverse reactions and was discharged

DISCUSSIONS

Wilms tumor (WT) is the most common type renal cancer in pediatric population. It affects approximately one child per 10,000 worldwide before the age of 15 years. The median age at diagnosis of WT is approximately 3.5 years.[11] Wilms tumor typically presents with non-specific clinical features such as abdominal pain (30%–40%), a palpable abdominal mass, hypertension (approximately 25%, often normalizing after nephrectomy), gross haematuria (12%–25%), unexplained fever, anorexia, and weight loss (seen in about 10% of cases).

Similar features were observed in our patient, who presented with abdominal swelling, vomiting, and reduced urine output.[12] More than 15 different syndromes are associated with WT, including WAGR, Denys-Drash and Beckwith-Wiedemann, only 10% of WT cases are associated with an underlying constitutional mutation, and therefore the etiology of most cases is unknown.[13] Wilms tumor is primarily evaluated using imaging modalities. Abdominal ultrasonography is the initial diagnostic investigation of choice to identify and confirm a renal mass. Cross-sectional imaging with contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) is considered the standard for further characterization, staging, and surgical planning. Echocardiography is indicated in selected cases to assess cardiac function and to evaluate for possible venous extension of tumor thrombus into the inferior vena cava and right atrium.[14] Multimodality treatment is necessary for the management of Wilms tumor which is as per SIOP (preop chemo-nephrectomy-adjuvant chemo ± radiotherapy) or NWTS protocol where nephrectomy is the primary treatment followed by adjuvant chemotherapy ± radiotherapy.[15] For patients with unilateral localized tumor, 4-week pretreatment with vincristine (weekly) and dactinomycin (biweekly) is given.[16] NWTSG favors surgery first while the SIOP advocates the use of neoadjuvant chemotherapy, the SIOP approach of preoperative chemotherapy makes surgery safer and achieves good survival results.[17] Childhood and adolescent cancer survivors require close monitoring because cancer therapy side effects may persist or develop months or years after treatment.[18]

CONCLUSION

Fetal Wilms tumor (WT) is an extremely rare condition, but advances in prenatal imaging have increased its detection. Its management is complex and requires a coordinated multidisciplinary approach involving pediatric radiology, oncology, surgery, and radiotherapy. Early diagnosis—especially when tumors are still small and localized—significantly improves survival outcomes and can be life-saving. Early detection and timely surgical intervention after birth significantly improve survival. Prognosis is generally excellent but is influenced by tumor stage, histology, and complications such as polyhydramnios. Identifying associated predisposition syndromes enables structured surveillance for related malignancies like hepatoblastoma and neuroblastoma, allowing earlier treatment, reduced therapy intensity, and better overall outcomes.

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