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Case Report
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Familial Metachronous Wilms' Tumour in a Sibling with Previous Bilateral Synchronous Wilms' Tumour: A Rare Familial Cluster

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Abstract

Wilms Tumour (WT) is the most common renal malignancy of childhood, typically occurring sporadically. Familial WT accounts for fewer than 2% of cases and often involves germline predisposition syndromes. Metachronous WT in siblings - particularly when one sibling previously presented with bilateral synchronous disease - is exceptionally rare. We report a case of a child presenting with unilateral WT whose older sibling had been treated several years earlier for bilateral synchronous WT. This case highlights the importance of familial risk assessment, genetic counselling, structured surveillance, and early detection strategies in families with suspected hereditary WT predisposition.

Keywords: Wilms Tumour; Familial Wilms Tumour; Bilateral Synchronous Wilms Tumour; Metachronous Tumour; Nephrogenic Rests; Hereditary Cancer Predisposition; Paediatric Renal Tumour; Genetic Counselling; Nephron-Sparing Surgery; Childhood Cancer Surveillance

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Introduction

Wilms tumour arises from aberrant nephrogenic rests and is associated with several genetic loci, including WT1, WT2, WTX, and genes involved in microRNA processing [1-7]. Bilateral synchronous WT occurs in approximately 5-10% of cases and is strongly associated with germline mutations [2, 8, 9]. Familial WT is rare, and metachronous presentation in siblings is even more unusual [1]. The presence of bilateral synchronous disease in one sibling significantly raises suspicion for an inherited predisposition, even in the absence of syndromic features [1, 3]. This case contributes to the limited literature on familial WT clusters and underscores the need for long-term surveillance protocols for at-risk siblings [1, 2, 10].

Case Report

Sibling 1 (Index Case)

A previously healthy child presented at age 3 with increasing abdominal distension. Examination revealed a firm bilateral flank mass. There were no dysmorphic features, aniridia, genitourinary anomalies, or developmental concerns.

Investigations

- Ultrasound: Bilateral heterogeneous renal masses with preserved intervening parenchyma. ; no metastases.in lymph nodes liver and chest radiograph was normal.
- Intravenous urography: Two large renal masses consistent with bilateral WT.
- Laboratory tests: Normal renal function; mild anemia.

Management

The child received neoadjuvant chemotherapy per SIOP protocol, followed by bilateral nephron-sparing surgery. Histology confirmed favorable-histology WT with nephrogenic rests. Postoperative chemotherapy was completed without complications.

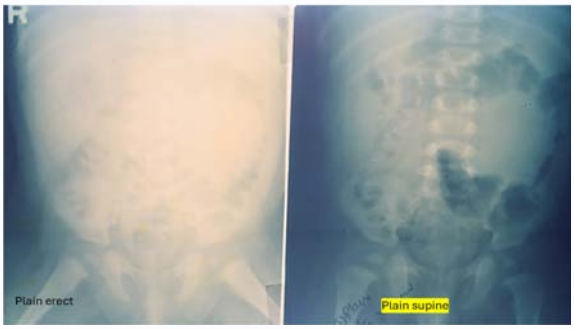

Figure 1: Plain erect and supine abdominal radiographs demonstrating a left-sided soft tissue mass in the lumbar region. The erect film reveals displacement of bowel loops and a subtle air–fluid level, while the supine view confirms the presence of a well-defined left renal mass. These findings are consistent with a unilateral Wilms tumour.

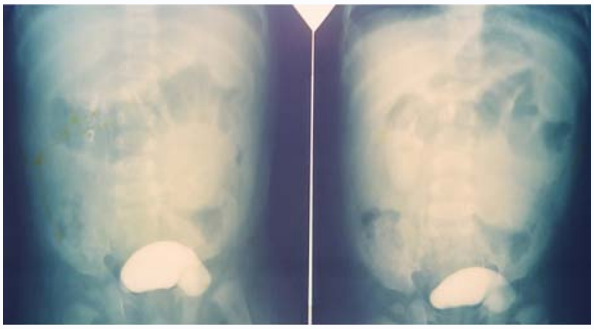

Figure 2: Intravenous urography showing prompt excretion of contrast on right side with delayed function and a large soft tissue mass at the lower pole of left kidney. Delayed film shows almost clearance from right kidney while left lower polar mass showing retention of contrast.

Outcome

The child remains in remission at 6-year follow-up with stable renal function.

Sibling 2 (Current Case)

A younger sibling, previously well, presented at age 4 with a short history of abdominal fullness noted by parents during bathing. There was no pain, hematuria, or weight loss.

Examination

- Palpable, non-tender left flank mass, No dysmorphic features, Normal blood pressure, No evidence of syndromic stigmata.

Investigations

- Laboratory tests: Normal renal function; LDH mildly elevated.
- Chest Radiograph: No pulmonary metastases. Abdominal erect and supine plain films showed a large sift tissue mass in left lumbar region displacing bowel loops (Figure 1).
- Ultrasound: Large left renal mass; right kidney normal. 8 cm heterogeneous mass arising from the left kidney, no vascular invasion, no contralateral lesions. No renal vein invasion.
- Intravenous urography showed normal functioning right kidney and left kidney confirmed large mass at the lower pole (Figure 2).

Genetic Evaluation

Given the family history, both siblings underwent genetic testing:

- WT1 sequencing: Negative.
- 11p15 methylation analysis: Normal.
- MicroRNA processing gene panel: Variant of Uncertain Significance (VUS) in DROSHA.

Although no pathogenic variant was identified, the familial pattern suggested a possible undetected germline predisposition.

Management

The child received preoperative chemotherapy, followed by left radical nephrectomy. Histology confirmed favorable-histology WT without anaplasia. Postoperative chemotherapy was completed per protocol.

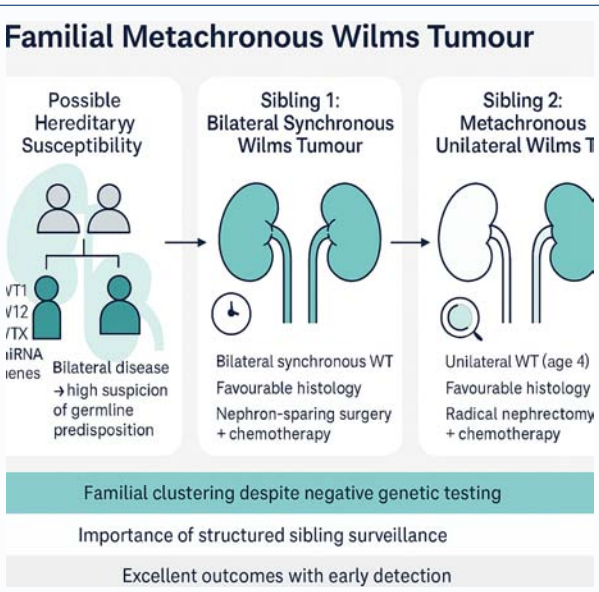

Figure 3: Graphical abstract illustrating familial clustering of Wilms tumour in two siblings. Panel 1 depicts possible hereditary susceptibility with a family tree and associated genetic loci (WT1, WT2, WTX, miRNA). Panel 2 shows Sibling 1 diagnosed at age 3 with bilateral synchronous Wilms tumour, treated with nephron-sparing surgery and chemotherapy. Panel 3 shows Sibling 2 diagnosed metachronously at age 4 with unilateral Wilms tumour, treated with radical nephrectomy and chemotherapy. The schematic highlights the importance of structured sibling surveillance and early detection in suspected familial cases.

Outcome

At 24-month follow-up, the child remains disease-free with normal function of the remaining kidney.

Differential Diagnosis included Wilms' tumour, Clear cell sarcoma of the kidney, Renal cell carcinoma (rare in children), Mesoblastic nephroma (unlikely at this age), Neuroblastoma (excluded by imaging characteristics and renal origin).

Treatment

Both siblings were treated according to contemporary SIOP protocols, with neoadjuvant chemotherapy, surgical resection, and adjuvant therapy tailored to stage and histology. Nephron preservation was prioritised in the sibling with bilateral disease.

Outcome and Follow-Up

Both siblings remain in remission. Renal function is stable.

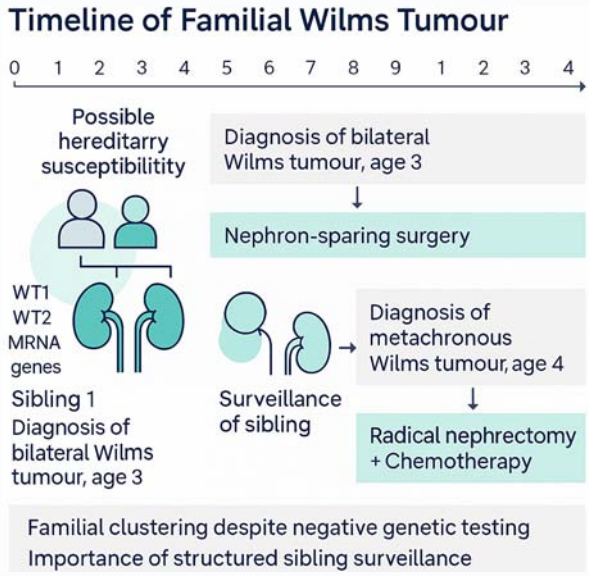

Figure 4: Timeline illustrating the familial presentation of Wilms tumour in two siblings. Sibling 1 was diagnosed at age 3 with bilateral synchronous Wilms tumour and treated with nephron-sparing surgery followed by chemotherapy. Following this diagnosis, structured surveillance was initiated for Sibling 2. One year later, Sibling 2 was diagnosed metachronously at age 4 with unilateral Wilms tumour and underwent radical nephrectomy and chemotherapy. The timeline highlights the importance of early surveillance in families with suspected hereditary predisposition.

Ongoing surveillance included:

- Ultrasound every 3-4 months for the first 2 years.
- Annual renal function monitoring.
- Long-term monitoring for chemotherapy-related late effects.

Discussion

This case describes an exceptionally rare familial pattern of Wilms' tumour: metachronous unilateral disease in one sibling occurring several years after the other sibling presented with bilateral synchronous Wilms' tumour. Familial Wilms' tumour accounts for fewer than 2% of cases [11], and the combination of bilateral synchronous disease in one child followed by metachronous disease in a sibling is scarcely reported in the literature. Our report highlights the clinical significance of structured surveillance, the challenges of genetic interpretation when no pathogenic variant is identified, and the importance of family-centred counselling in suspected hereditary tumour predisposition.

We believe this case will be of particular interest to clinicians in paediatric oncology, genetics, nephrology, and primary care, as it reinforces the need for vigilance in families with atypical or bilateral Wilms tumour presentations.

This case illustrates an exceptionally rare familial pattern of WT: metachronous unilateral WT in one sibling following bilateral synchronous WT in another.

Key considerations include:

Familial Risk

Siblings of children with WT have a higher risk than the general population, particularly when the index case has: Bilateral disease, Early age of onset, Nephrogenic rests, Suggestive family history [12].

Genetic Predisposition

Although no pathogenic variant was identified, the presence of: Bilateral synchronous WT in one sibling, Metachronous WT in another strongly suggests an underlying hereditary predisposition, possibly involving genes not yet fully characterised [13].

Surveillance

Early detection significantly improves outcomes. Current recommendations for at-risk siblings include:

Regular abdominal ultrasound from birth or time of index diagnosis, continued surveillance until at least age 7-8 and Consideration of extended monitoring when familial clustering is present [14].

Clinical Implications

This case reinforces the need for: Multidisciplinary management, Genetic counselling, Family-centred surveillance strategies, Awareness of non-syndromic hereditary WT patterns.

Patient's Perspective

"When our first child was diagnosed with Wilms' tumour, our whole world changed overnight. We learned to live with hospital appointments, scans, and the constant worry of what each result might show. When treatment finished, we tried to rebuild a sense of normality, always hoping the worst was behind us.

Years later, when our younger child began to show similar symptoms, we recognised the signs immediately. It was frightening to face the possibility of another tumour in the family, but having been through the process once, we also knew the importance of acting quickly. The early scans confirmed our fears, yet we felt more prepared this time—more aware of the treatments, the teams involved, and the strength our family had already shown.

Although the genetic tests did not give us clear answers, we are grateful that both children received timely care and are now doing well. We have learned the value of surveillance and of trusting our instincts as parents. Our hope is that sharing our story will help other families feel less alone and encourage early assessment when something doesn't feel right."

Learning Points

- Familial Wilms' tumour is rare, and metachronous disease in siblings is exceptionally uncommon.
- Bilateral synchronous WT in one sibling should prompt genetic evaluation and structured surveillance of all siblings.
- Absence of a detectable pathogenic variant does not exclude hereditary predisposition.
- Early detection through surveillance enables curative treatment with organ preservation where possible.
- Long-term follow-up is essential to monitor renal function and late effects of therapy.

Conclusion

This familial cluster - metachronous unilateral Wilms' tumour in one sibling following bilateral synchronous Wilms tumour in another - highlights an exceptionally rare pattern of pediatric renal malignancy. Even in the absence of an identifiable pathogenic variant, the presentation strongly suggests an underlying hereditary

predisposition. This case reinforces the importance of maintaining a high index of suspicion for familial Wilms' tumour when bilateral disease occurs in an index case, and it underscores the value of structured surveillance for at-risk siblings. Early detection enabled timely, curative treatment in both children, with preservation of renal function and favorable outcomes. As genomic understanding evolves, such cases emphasise the need for continued refinement of genetic testing panels, long-term follow-up strategies, and family-centred counselling to optimise care for children with potential hereditary cancer risk.

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