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Case Report

Association of Wilms Tumor with Crossed Fused Renal Ectopia in Children: A Case Report

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Abstract

Introduction: Wilms tumor is the most common renal malignancy in children. The occurrence of Wilms tumor with various congenital genitourinary anomalies has been reported, particularly in horseshoe kidneys, hypospadias, disorder of sexual development, and double collecting system. However, Wilms tumor with crossed renal ectopia is a rare finding. **Case Description:** We are reporting a case report of Wilms tumor in a 3-year-old girl who presented with a huge left flank mass with cross-fused renal ectopia. After the initial workup and triphasic computed tomography scan of the chest, abdomen, and pelvis for confirmation of diagnosis and metastasis, the patient underwent image-guided tissue biopsy, followed by neoadjuvant chemotherapy, left radical nephrectomy with separation of fused right ectopic renal moiety, and adjuvant chemoradiation. **Practical Implication:** This report shows an association of Wilms tumor with cross-fused renal ectopia, a rare combination. Unusual findings on imaging with unilateral flank mass should be considered as part of the differential diagnosis for this rare finding. Adjuvant chemotherapy and modern imaging helped delineate the anatomy and ease the surgery for safe resection, improving the overall outcome.

Keywords: Radical nephrectomy, renal ectopia, Wilms tumor

Introduction

The occurrence of Wilms tumor with various congenital anomalies is well known.[1] Crossed fused renal ectopia is a rare congenital malformation wherein both kidneys are present unilaterally and fused. The incidence of crossed-fused renal ectopia has been reported to be approximately 1:1000 live births. It has varied presentations, including nephrolithiasis, pelvic ureteric junction obstruction, cystic dysplasia, and renal tumors.[2]

The pre-operative diagnosis of tumors in children has paramount importance in the presence of this rare anomaly. It should be differentiated and investigated due to complex anatomy and disease-specific treatment for renal malignancy. There were only two cases of renal ectopia with Wilms tumor reported earlier around the world in children, and few of them are reported in the adult population. We are reporting a case of a 3-year-old girl who underwent left radical nephrectomy with complete separation of mass from the right renal ectopia after

confirmation of diagnosis on image-guided biopsy and neoadjuvant chemotherapy.

Case Description

A 3-year-old female child presented with abdominal mass and gradual distension for 2 months. Her medical and family history was insignificant. Physical examination revealed an active, irritable child weighing 13 kg. The abdomen was soft-distended, with a non-tender palpable mass at the left flank and pelvis, crossing midline without any lymphadenopathy. The rest of the systemic examination was unremarkable.

Complete blood count, chemistry profile, urinalysis and 24-h urinary vanillylmandelic acid, and chest radiograph were unremarkable. Ultrasound and tri-phasic computed tomography (CT) scan of the abdomen and pelvis were suggestive of the ectopic position of the right kidney in front of the lumbosacral junction with a huge retroperitoneal left hemi abdomen mass measuring 14 cm by 13 cm. The tumor arose from the left renal moiety compatible with a large Wilms tumor with multiple bi-lobar metastatic hepatic lesions [Figure 1]. Technetium-99m diethylene triamine penta-acetic acid (Tc-99m DTPA) showed the estimated glomerular filtration rate of 80 ml/min. An ultrasound-guided trucut biopsy of the renal mass revealed spindle cell neoplasm with the possible differentials of clear cell sarcoma and a mesenchymal component of Wilms tumor.

Diagnosis and management

After the initial workup and diagnosis, patient was discussed in a multidisciplinary tumor board and given neoadjuvant chemotherapy – actinomycin, vincristine, and doxorubicin for six cycles according to Children's Cancer and Leukemia (CCLG) Umbrella Protocol followed by reassessment scan and surgery for local control.[3] A reassessment scan showed a significant response to chemotherapy with a decreased residual tumor size of 10 cm by 9 cm [Figure 2].

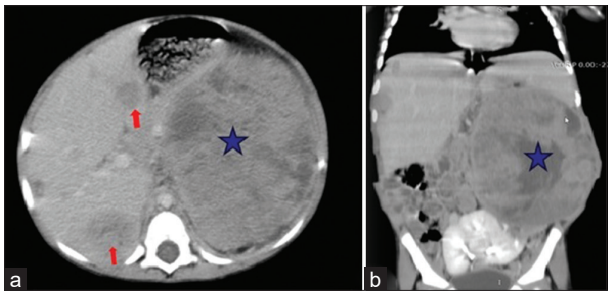

Figure 1: (a) Baseline axial view of contrast computed tomography scan showing huge retroperitoneal mass (star) arising from the left renal moiety with multiple bi-lobar liver metastatic lesions (arrowhead) and (b) coronal view showing left renal mass and normal ectopic right kidney

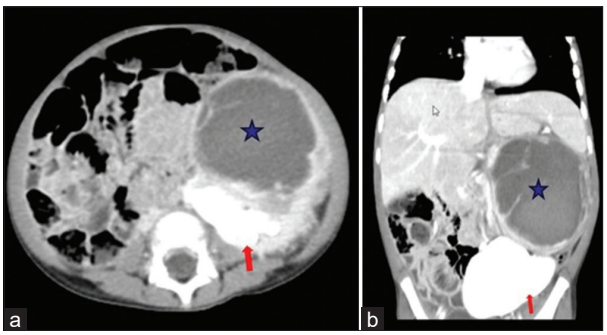

Figure 2: (a and b) Re-assessment axial and coronal view of contrast computed tomography scan showing significant response to chemotherapy and reduced size of the left renal moiety mass (star) and normal ectopic right kidney

The patient was explored through the left supraumbilical transverse incision, and after colonic mobilization left renal mass was seen adhered to the tail of the pancreas, spleen, and diaphragm. On further dissection, crossed fused renal ectopia with the right ectopic kidney attached to the lower pole of the left renal mass was identified, and the collecting system of the left kidney (mass) was draining into the pelvis of the right kidney with the duplex system. However, separate pelvis, ureter, and vasculature of the right kidney were identified and well preserved. Hence, the mass was completely resected with clear margins after ligation of renal vessels and ureter without complication. The child remained well postoperatively and was discharged on the 3rd post-operative day.

Histopathology revealed residual Wilms tumor with more than 90% therapy-induced necrosis and remaining epithelial and blastemal components [Figure 3]. On risk stratification, it was metastatic stage IV Wilms tumor, local disease stage I with low-risk group A and given adjuvant chemotherapy for 2 weeks followed by reassessment scan for surgical control of liver metastasis. On the follow-up scan, there was progressive disease (increase in the size and number of hepatic lesions), and treatment escalated to high-risk chemotherapy (cyclophosphamide, doxorubicin, and etoposide) for four cycles and planned radiotherapy (External beam radiation therapy) for liver metastasis. However, after 4 months of follow-up, the parents did not continue further treatment, and the patient was lost to follow-up.

Discussion

Wilms tumor (nephroblastoma) is the most common pediatric renal mass, accounting for almost 90% of all renal masses and representing 7% of all malignant tumors in children.[4] Wilms' tumor has been associated with several syndromes, including WAGR syndrome, Beckwith–Wiedemann syndrome, Denys–Drash syndrome, and Edwards or Perlman syndrome.[5] The association of Wilms tumor with genitourinary anomalies like horseshoe kidneys is well known in the literature.[1,6] However, the occurrence of Wilms tumor in orthotopic kidneys with cross-fused renal ectopia is reported as outstandingly rare in children.

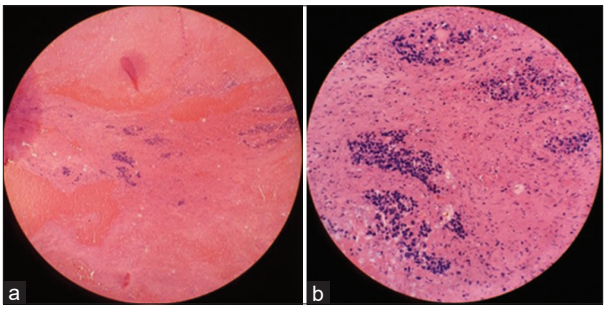

Figure 3: (a and b) Histopathology revealed a residual Wilms tumor with epithelial and blastemal components and more than 90% therapy-induced necrosis with clear margins

In the literature review, Berant *et al.*, in 1975, reported the first case of Wilms tumor in the crossed ectopic kidney, which was diagnosed with the help of intravenous pyelography and successfully treated by excisional biopsy and chemoradiation.[7] In the second case of Wilms tumor in crossed fused renal ectopia reported by Redman in 1977, excretory urography demonstrated crossed renal ectopia, and aortography confirmed the presence of a tumor on the right side of the abdomen.[8]

With the advancement of a multidisciplinary team approach, the survival rate for Wilms tumor approaches 90%.[9] Therefore, a careful pre-operative diagnosis and planning for surgery is necessitated in the successful management of Wilms tumor associated with this rare anomaly. Crossed renal ectopia may present as an abdominal mass due to renal infection, obstruction leading to hydronephrosis, or calculus formation. On investigation, the apparent anatomic abnormality, like crossed renal ectopia, may overshadow the possible presence of a mass, which will thus be overlooked if not specifically sought.

In our case, the pre-operative chemotherapy provided a significant response and decreased the tumor size. Furthermore, tri-phasic CT helped to delineate the renal vasculature and drainage of renal units for complete resection of the tumor, preservation of the uninvolved renal unit, and avoiding unpredicted complications. To the best of our knowledge, this will be the third case report in the literature of nephroblastoma in orthotopic kidneys with cross-fused renal ectopia, unlike the previous two cases described have a tumor in the ectopic kidney and successfully treated with multimodal therapy.

The association of Wilms tumor with crossed renal ectopia is confirmed to be a rare entity. Unusual findings on imaging with unilateral flank mass should be considered as part of the differential diagnosis for this rare finding. Adjuvant chemotherapy and modern imaging can improve surgical outcomes by providing accurate staging and clearer anatomy.

Acknowledgment

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Author Contributions

Conceived and designed the analysis: SA, TL; Collected the data: SA, MBS; Contributed data or analysis tools: MAS, MBS; Performed the analysis: TL; Wrote the paper: SA, TL, MAS.