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Photo Clinic

Diffuse Bilateral Nephroblastomatosis: A Pediatric Oncologic Challenge

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A 12-month-old boy was brought to the emergency department for a 2-week history of irritability and abdominal enlargement. The only positive finding on physical examination was abdominal distension. Initial imaging included an abdominal ultrasound which showed bilateral nephromegaly along with multiple bilateral hypoechoic lesions scattered within the renal parenchyma. Spiral abdominopelvic CT-scan demonstrated bilateral renal enlargement and loss of normal architecture with renal parenchyma completely replaced by homogenous low attenuating peripheral masses (Figure 1). Based on the radiological features, bilateral nephroblastomatosis was suggested for the patient. The patient underwent an open renal biopsy (wedge biopsy) to rule out wilms tumor. The pathology revealed sheets and nests of atypical cells with high nuclear/cytoplasmic ratio and vesicular nuclei with moderate mitotic activity. Small numbers of rosettes, trabecular structures and abortive glomeruli were also observed. There was no evidence of normal renal tissue. The pathologist could not definitely rule out Wilms tumor based on the results of the wedge biopsy. IHC study for WT1 showed strong nuclear positivity. According to the radio-pathological findings, the histopathology was reported as "nephroblastic process" (nephrogenic rest). In order to characterize the exact pathology of the kidneys, contrast-enhanced MRI was performed in which T1/T2 weighted images showed complete replacement of renal parenchymas with thick rind of hypointense masses with no obvious enhancement compatible with diffuse nephroblastomatosis (Figures 2).

Nephrogenic rests (NRs) are nonencapsulated structures of persistent immature renal tissue with histological features resembling Wilms tumor. Multiple or diffuse NRs are known as nephroblastomatosis (NB). NB; a premalignant lesion, represents the persistence of nephrogenic blastema beyond the 36th week of gestation. NRs frequently either regress or differentiate; however, they can become malignant if they persist. NRs are seen in 1% of the general population, in the adjacent renal parenchyma of about 40% of patients with unilateral wilms tumors and in almost 100% of bilateral tumors. It is recommended to monitor closely patients with NR, since they are associated with an increased risk of development of Wilms tumor.[1] In 1990 Beckwith and colleagues proposed classify NB into four categories based on their location: The perilobar (PLNB), intralobar (ILNB), combined and universal (panlobar) or diffuse in 1% of cases.[2, 3] Nephrogenic rests are further classified histologically as dormant, sclerotic, hyperplastic, and neoplastic. Neoplastic nephrogenic rests are equivalent to wilms tumor and represent clonal proliferation.[4] In diffuse hyperplastic perilobar nephroblastomatosis (DHPLN), the cortical surface of one or both kidneys is composed of hyperplastic nephroblastic tissue.

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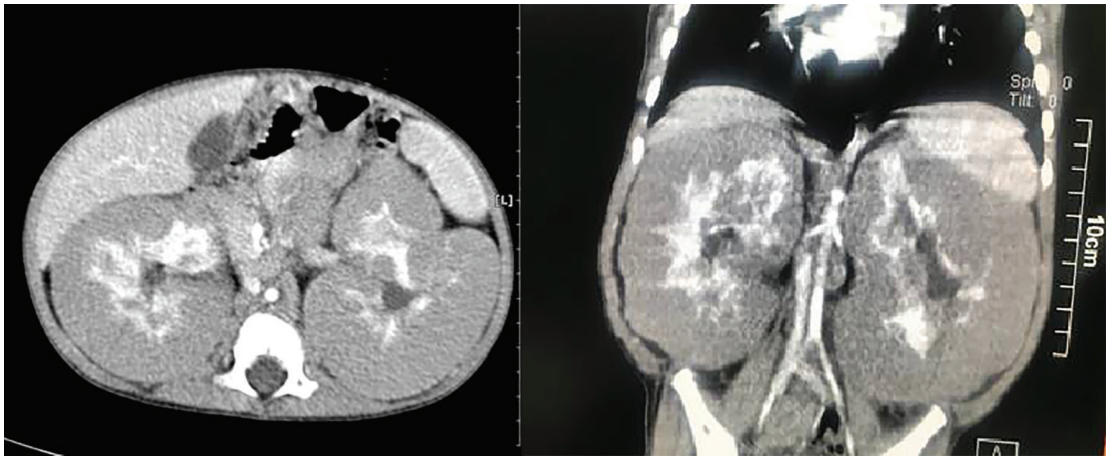

Figure 1: Abdominopelvic CT-scan shows bilateral renal enlargement and loss of normal architecture with renal parenchyma completely replaced by homogeneous low attenuating peripheral masses.

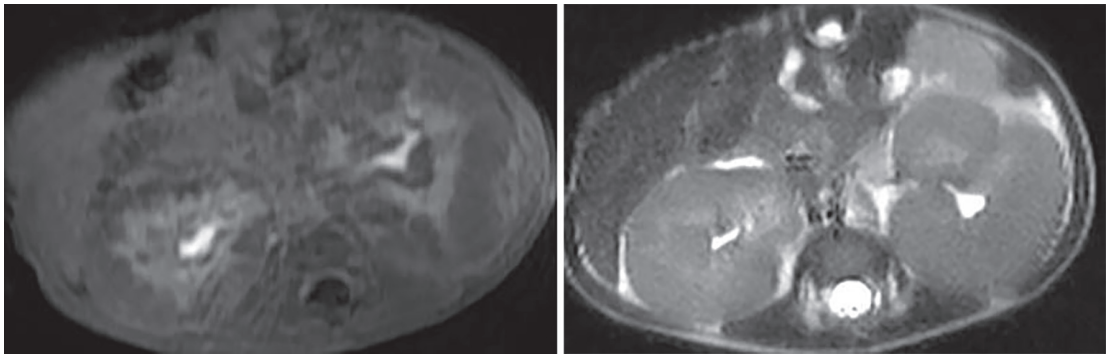

Figure 2: T1 and T2 weighted MRI images show complete replacement of renal parenchymas with thick rind of hypointense masses with no obvious enhancement compatible with diffuse nephroblastomatosis.

Radiographically, DHPLN is characterized by massive enlargement of the kidney with a rind-like expansion of the renal cortex of homogeneous signal intensity and preservation of the renal shape.[5] Histological distinction between Wilms tumor and nephrogenic rests is not always possible based on morphology alone and implementation of new molecular genetic tools may aid in this regard.[6] Fine-needle aspiration cytology is of limited value and is not able to distinguish nephroblastomatosis from wilms tumor.[7] Interestingly, in diffuse NB, the reniform contour of the kidneys is preserved with a thick rind like peripheral tissue.[8] This is in contrast to wilms tumor that appears as a spherical mass on imaging.

Therapeutic management of nephroblastomatosis is debated. Preoperative chemotherapy is suggested to preserve normal parenchyma and to prevent transformation of nephroblastomatosis into Wilms tumor. Surgery is not recommended as the initial treatment for nephroblastomatosis and should be avoided due to the increased risk of a tumor developing in the contralateral kidney.[7]

The entity of "diffuse nephroblastomatosis" should be kept in mind of pediatric oncologists; next to "Wilms tumor", and followed carefully by high-resolution imaging techniques and in case of any radiologic change, pathologic assessment is recommended to ensure that the appropriate measurement is taken.

Conflict of Interest: None declared.

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