BILATERAL WILMS TUMOUR AND NEPHROBLASTOMATOSIS
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INTRODUCTION:
Wilm’s tumour (nephroblastoma) is the most common malignant pediatric renal tumours accounting to about 85% kidney related mass lesions and 6% of all childhood cancers. It typically occurs in early childhood (1 - 11 years) with peak incidence between 3 and 4 years of age. Approximately 80% of these tumours are found before the age of 5 years. They occur even earlier, typically between 2 and 24 months of age whilst being associated with some congenital anomalies such as cryptorchidism (2.8% of cases), hemi hypertrophy (2.5%), hypospadias (1.8%), and sporadic aniridia (4). We report a case with bilateral perilobar nephrogenic rests and Wilm’s tumour without any associated syndrome.

CASE REPORT:
A 4 years-old male child came with progressive increase in abdominal girth, with associated loss of weight and history of irregular fever in the past. Clinical examination revealed a hard abdominal mass.

IMAGING FINDINGS:
On ultrasonography bilateral heteroechoic well defined lobulated mass with nodular calcifications within were detected in the upper pole of both Kidneys. In addition there were few hypoechoic lesions (approximately 2cm diameter) also noted distributed in both kidneys. Plain and post intravenous contrast abdomino-pelvic imaging was performed with Siemens Somatom Sensation 64 (0.8-mm section thickness, pitch of 1.5, 117mAs, and 120 kV) and 25ml of non ionic Omnipaque as intravenous contrast medium. C. T. imaging revealed enlarged kidneys (12.9 x 12.3 x 6.6 and 12.1 x 11.9 x 6.4cms), with large well defined heterogenous masses in soft tissue density seen bilaterally (24 HU) displacing the liver and spleen. Some calcific foci were also noted within however no evidence of any hemorrhage detected. Heterogeneous enhancement was noted on administration of intra venous contrast with displacement of IVC to left side. The renal artery and vein had normal appearances. No evidence of any invasion into the renal vein or inferior vena cava seen. Delayed imaging demonstrated no definite continuity of the mass with the collecting system. However multiple sub capsular, low-attenuating (18 HU), nodular lesions, about 1.5 cm in diameter, were seen in both kidneys; in the mid and lower poles suggestive of nephrogenic rests. The lesion showed poor enhancement on post contrast.

DISCUSSION:
Bilateral large renal mass in the age group of early childhood (3-4yrs) the possibility could be Wilms tumor or neuroblastoma. The lesions are well defined and not illdefined with well defined margins with absence of poorly defined margins. The lesion also shows displacement of IVC and not encasing the vessels with absence of extension into chest and elevation of aorta. With these findings a diagnosis of bilateral Wilm’s tumor with nephroblastomatosis was offered which was confirmed subsequently on histopathological examination. Wilm’s tumor (nephroblastoma) is a common pediatric tumor, with peak incidence at 3-4 years of age (5) and accounts for 87% of pediatric renal masses. The incidence is approximately 1:10,000 populations (1, 2). Wilm’s tumor is bilateral in 4%–13% of children (4). Metanephric blastema that persists at birth is called a nephrogenic rest; multiple nephrogenic rests are referred to as nephroblastomatosis. According to autopsy studies, evidence of nephrogenic rests is present in approximately 1% of infants. Malignant transformation of nephrogenic rest is likely to occur in less than 1% of these affected children. Nephroblastomatosis are often suggested as premalignant condition (6). Nephrogenic rests are classified into two groups: (a) intra lobar, occurring anywhere within the lobe and (b) perilobar, occurring in the cortex, covering the lobe. Perilobar rests are more common than intralobar rests and usually are multiple and well circumscribed. Intralobar rests are irregular and usually occur singly or in a small number (7). Perilobar rests are associated with Beckwith-Wiedemann syndrome (gigantism, macroglossia, omphalocoele, genitourinary anomalies) and hemi hypertrophy. Intralobar rests are associated with Drash syndrome (ambiguous genitalia and progressive renal failure in boys) and sporadic aniridia; Wilm’s tumor is more commonly associated with intralobar rests than with perilobar...
rests (5, 6). Wilm’s tumor is associated with congenital anomalies such as cryptorchidism (2.8% of cases), hemi hypertrophy (2.5%), hypospadias (1.8%), and sporadic aniridia (4). Bilateral perilobar nephrogenic rests and Wilm’s tumor, without any associated syndrome is rare. Treatment is generally with surgical management and chemotherapy.

REFERENCES:

Figures:
Figure 1 (a)

![Image](image1_a)

Figure 2 (a)

![Image](image2_a)

Figure 2 (b)

![Image](image2_b)

Legends:
Fig. 1a Pre contrast C.T. Images in axial section show bilateral large renal mass in upper pole & mid pole (arrow) and multiple nephroblastomatosis (arrow head).
Fig. 1b CT axial section demonstrates calcification within renal mass (arrows).
Fig. 2a Post contrast C.T. Images in coronal & axial, depicting the heterogenous pattern of enhancement of the tumor
Fig. 2b C.T. axial and coronal section showing multiple nephroblastomatosis (arrow).