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Wilms tumor in Newborn: A Case Report and Update

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Abstract: *Wilms tumor (WT) in a children younger than 3 months is a very rare condition comprising less than 2% of cases. Here we present arare case of WT diagnosed at the age of only 1 month. A female newborn baby when came for 1st visit immediate after birth to a pediatrician, a mass was noted in left lumber region . Her first USG report was not conclusive but repeat USG at 1 month showed focal lesion within left kidney . CT scan also revealed mass lesion suggestive of WT.Total nephrectomy was performed and histologic examination confirmed WT. Patient received postoperative chemotherapy with vincristine and actinomycim-D. Now she is on regular follow up for one and a half years after completion of treatment without any complications and signs of recurrence.*

Key Words: *Newborn, abdominal mass, Wilms tumor*

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I. Introduction

Wilms tumor is seldom seen in a neonate and prenatal diagnosis is rare.[1]It is the most frequent renal neoplasm, comprising 95% of renal tumors in children under 15 years and approximately 7% of all pediatric neoplasms. In 80% cases, it affects children under 5 years.[2] In infants younger than 3 months, it is a very rare condition, comprising less than 2% of cases.[3] Neonatal WT represented 0.11% of all patients included in clinical trials of the National Wilm's Tumor Study.[4]About 15% of children with WT have associated congenital anomalies like aniridia (1.1%), hemihypertrophy (2.9%), musculo-skeletal (2.9%) and genito- urinary (4.4%) anomalies.[5] Abdominal palpable mass and enlargement of abdominal girth are the most common presentations of renal tumors (77%).Other symptoms are abdominal pain, fatigue and vomiting in about 30% of patients, hypertension , fever or hematuriain 25% and more rarely other symptoms like varicocele, hepatomegaly, ascites, congestive heart failure and acquired Von Willebrand disease that can occur mainly as aresult of the presence of intra-atrial, renal vein or inferior vena cavaltumors.[4] Examination usually reveals a large smooth, non-tender, immobile and firm abdominal mass that does not cross the midline.[6]An antenatally detected renal tumor may be associated with polyhydramnios, hydropsfetalis and acute foetal distress.[7]This study aims to report a case of a newborn who presented with an abdominal mass just after birth and was diagnosed as a left sided Wilms tumor at only one month of age.

II. Case Summary

A female baby was born by lower uterine cesarean section (LUCS) at 38 weeks of gestation at Ad-din Women's Medical College Hospital due topremature rupture of membrane(PROM) with uneventful perinatal period . Her mother was on regular checkup and all antenatal USGswere normal.When she was visited by a paediatrician for first time after birth, a mass was noted on left lumber region measuring about 4 x3 cm in diameter. She was otherwise healthy having birth weight 2.5 kg & length 50 cm with normal reflex and activity.

Her blood pressure (BP) could not be recorded, but other vital signs were within normal limit. She did not have any ocular, genitourinary and musculoskelital abnormalities

The baby was advised for ultrasonography (USG) of whole abdomen that revealed enlarged left kidney measuring about 5.6 x3.2 cm with loss of normal parenchymal echopattern,showing increased echogenicity of the cortices & poor differentiation between cortex & medulla with perinephric fluid collection. Then the baby was referred to an urologistand second USG of whole abdomen was suggested one month later that showed enlarged left kidney measuring about 6 x4.7 cm. A well defined fairly large mixed echogenic focal lesion was seen in the mid part of left kidney which was extended beyond the surface margin suggestive ofleft renal neoplasm. Then CT scan of whole abdomen was performed that showed strong heterogeneously enhancing well defined mixed density mass lesion measuring about 7.2 x4.5 x3.9 cm , noted in the left side of the abdomen arising from the upper and mid pole of the left kidney. The mass was displacing the left renal vesssels and bowel loops medially.Remaining part of left kidney appeared normal with normal excretion of the contrast media. All findings of CT scan were suggestive of nephroblastoma (left) . Her complete blood count(CBC) revealed-total count(TC) 10,000 /cmm, neutrophil(N)- 58.5 % ,lymphocyte(L)- 34.9 %, hemoglobin(Hb) 12.4 g/dl , platelets 3,51,000 /cmm. Her creatinine was 0.7 mg /dl (N), blood urea nitrogen - 8.12 mg /dl (N). Serum electrolytes ,urine routine and microscopic examination(R/M/E) and culture & sensitivity(C/S), liver function test, blood C/S , chest x ray(CXR) all were normal. So, our clinical and radiological diagnosis was Wilms tumor.

She underwent left nephrectomy at the age of 1 month 12 days under general anesthesia by an urologist. She had no post operative complications. Her biopsy specimen consisted of a nodular piece of tumor tissue measuring 6x5x3.5 cm on gross examination.The cut surface was homogeneous and gray white. On microscopic examination, section showed a Wilms tumor composed of blastema, stromal and epithelial cell components. The epithelial components formed abortive tubules and glomeruli. After 10 days of surgery, chemotherapy was started with Vincristine and Actinomycin-D. Total 19 cycles of chemotherapy were given over a period of four and a half months.After completion of treatment, the baby was on regular follow up initially 3 months interval for two times then 6 monthly. She was clinically evaluated and few routine investigations were performed like CBC, liver function test, USG of whole abdomen, serumcreatinine, CXR. Follow up will be continued up to 5 yrs. Now the baby is 25 months, her BP- 90/60 mm Hg, systolic - on 25th centile and diastolic- on 10th centile. Her weight(W)- 9.9 kg, Height(H)-82 cm, occipitofrontal circumference(OFC)-45.5 cm. Baby's W/H falls on 10thcentile, H/A- just below the 25th centile, OFC- on 5th centile. Now she is doing well without any complications and signs of relapse.

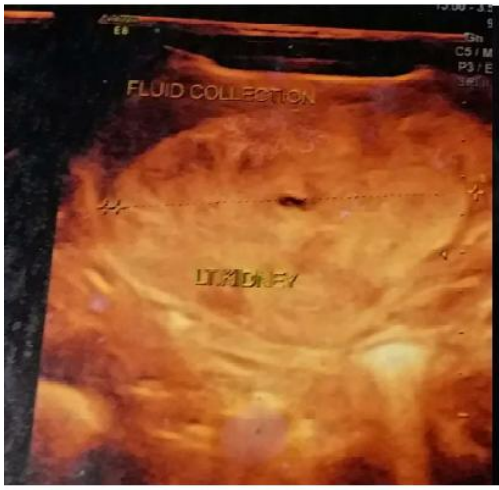

Fig 1: USG on 1st day showing enlarged left kidney with loss of corticomedullary differentiation and fluid collection.

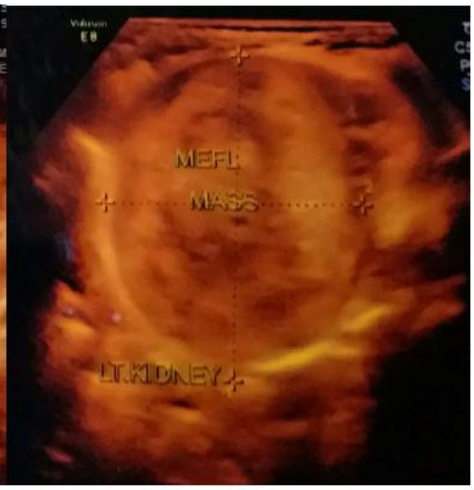

Fig-2: USG at 1 month showing well defined mixed echogenic focal lesion in the mid part of left kidney suggestive of left renal neoplasm.

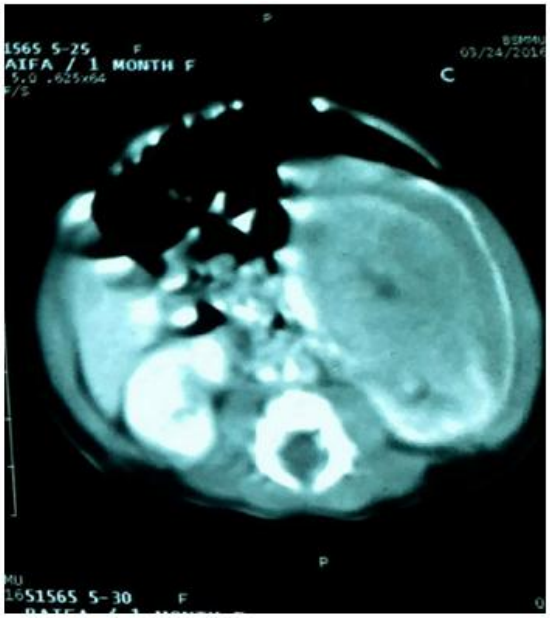

Fig.3: CT scan showing heterogeneously enhancing well defined mixed density mass lesion arising from upper &mid pole of the left kidney suggestive of nephroblastoma (left).

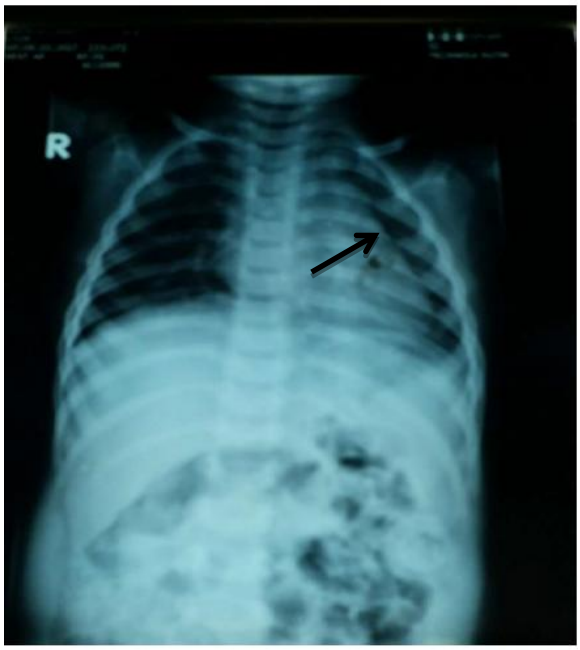

Fig.4: Normal CXR at 1 month

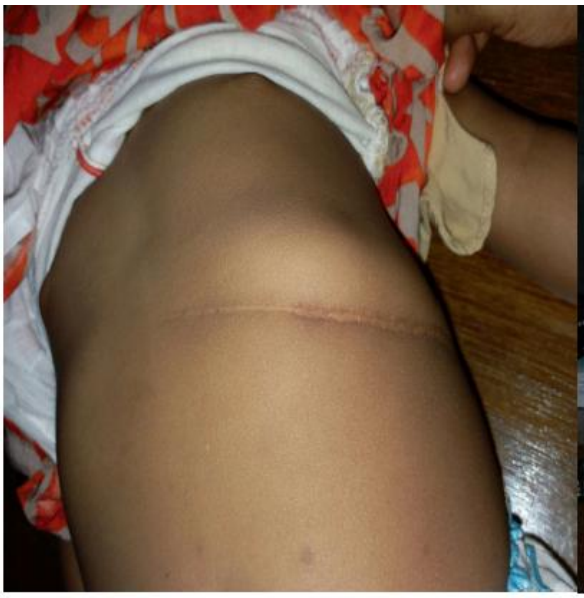

Fig. 5: Incision mark after total nephrectomy.

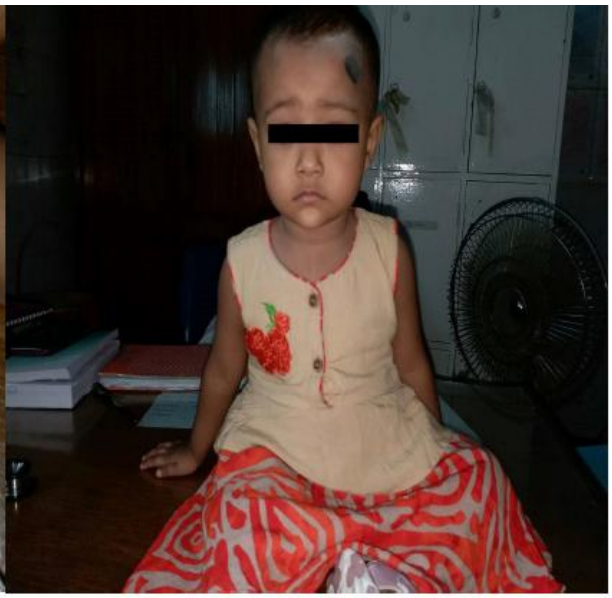

Fig. 6: Baby at the age of one and half year.

III. Discussion

Our case report describes a female patient who presented with abdominal mass just after birth and whose diagnosis was made as Wilms tumor only at one month. In the existing literature, very few case reports were published on WT at neonatal period. GenerallyWT is considered as a preschool children disease with median age of 3-5 years at the time of diagnosis. Our patient presented with palpable abdominal mass that is the most common presentation of renal tumors (77%).[4]

WT is primarily a sporadic disease and only 1 to 2 percent of individuals with WT have a positive family history.[8] The tumor usually affects a single kidney, but approximately 5 to 10 percent of children with WT have bilateral involvement.[9]Our patient presented with unilateral involvement. In some children, WT occurs as a part of a multiple malformation syndrome.[10] These syndromes include WAGR, Denys-Drash, and Beckwith-Wiedemann syndromes. Patients with associated congenital anomalies, such as aniridia or genitourinary abnormalities are also diagnosed at an earlier age.[11]

Congenital mesoblasticnephroma (CMN) is the most common renal solid tumor of the newborn period.[12]The main differential diagnoses of WT are neuroblastoma, mesoblasticnephroma, renal adenocarcinoma, rhabdomyosarcoma, hepatoblastoma, hepatocellular carcinoma, polycystic kidneys and hydronephrosis.[13]

Modern imaging techniques are valuable for diagnosis of WT before confirmation by histology.[14] Ultrasound with Color Doppler, computed tomography (CT) scan and magnetic resonance imaging (MRI) are all modern imaging techniques that have proved useful for diagnosis. Conventional ultrasound is the most common method for initial diagnosis of WT due to its non-invasiveness, affordability and availability.[15]Most of the causes of abdominal distension in children can be differentiated from WTusing ultrasound scan.[16] Neuroblastoma commonly crosses the midline and usually calcifies. Since it is a suprarenal tumor, it displaces the kidney while WT destroys and distorts the calyces because it is intrarenal.[17] In hydronephrosis, the entire collecting system is outlined as a series of connected fluid-filled channels. When only part of the renal collecting system is dilated, the condition may superficially resemble a tumor on sonographic examination, but close scrutiny can easily differentiate the 2 conditions.In Polycystic kidneys, ultrasonography shows diffusely enlarged kidneys with a generalized increase in echoes and the renal borders are poorly defined and corticomedullary differentiation is lost.[16] In our patient, USG of whole abdomen showed a well-defined large mixed echogenic focal lesion that arose from mid part of left kidney thereby excluding all other causes of abdominal mass. However, USG has poor cross-sectional anatomical information and it is less accurate in tumor staging.[18]

Cross sectional imaging like CT scan and MRI have been shown to be superior to ultrasonography.[19] CT has been found to have accuracy of 82% and capable of an explicit report in 89% of all cases.[20]CT is currently the technique of choice in the diagnosis and staging of renal masses in children, and is particularly relevant in recognition of size and site of lesion and densitometric patterns. It also provides an excellent visualisation of contiguous structures like vessels and lymph nodes.[19]On CT, Wilms tumor usually appears as a bulky, spherical intrarenal mass, very often with a well-defined rim of compressed renal parenchyma or

pseudocapsule surrounding it. Some tumors that arise from the periphery of the cortex may grow in an exophytic manner with bulk of the tumor seen outside the renal cortex.[21] In general, a heterogenous mass replacing the kidney and displacing adjacent organs can be demonstrated. The tumor is hypodense compared to the normal renal parenchyma on contrast enhanced CT scans with the areas of low attenuation coinciding with tumor necrosis, fat deposition or both.[22]CT scan of abdomen of our patient also demonstrated the similar findings.

WT arises anywhere in the kidney as embryologic precursors to renal cells. These cells mimic normal development of the kidney and consist of three components: blastema, epithelium, and stroma.[23] WT are grouped into 2 major types based on their histology- favorable and unfavorable histology. A favorable histology consists of well-differentiated cells, or cells that appear closely related to normal cell sources such as those mentioned [24] unfavorable outcomes are typically related to the anaplastic cell with diffuse or focal involvement, making up 10% of WT cases. The anaplastic cells are undifferentiated, which means they appear so abnormal, their origin cannot be determined. These cell types are more aggressive in their growth and behavior. Diffuse anaplasia bears theworst prognosis .[25] Our patient's histology was luckily favorable that consisted of blastema, epithelium and stroma. WT usually spreads into renal sinuses, intrarenal lymphatics and blood vessels.[26] Common sites for metastasis are the lungs, regionallymph nodes, and liver.[9]There was no evidence of metastasis in our case.

Prenatal detection of Wilms tumor is extremely rare.[27] Besides ultrasonography, foetalmagnatic resonance imaging (MRI) has been found to be helpful in resolving the diagnostic dilemma of foetal renal masses.[28] Imaging guided FNAC, when feasible can confirm the diagnosis.[29]Unfortunately, there is not much scope of prenatal intervention for foetal renal tumors and definitive intervention is usually deferred until birth.[30]However, in this case mother's antenatal USGs did not reveal any abnormalities.

Sarin et al presented a case of antenatally diagnosed left sided WT with features of hydropsfetalis in a girl baby.[30]Talini et al presented a case of nephroblastoma diagnosed at the age of 70 days .Mother noticed increased abdominal volume at 15 days of life with progressive worsening.[31]Starcevic et al reported on a boy with aniridia, cryptorchidism and facial dysmorphism recognized as WAGR syndrome (Wilms tumor, aniridia, genitourinary malformations and mental retardation) in neonatal period, subsequently confirmed by genetic testing. WT developed at the age of one year. [32]Choudhury et al published a case report on a 19 month old girl who was diagnosed to have WT and bilateral sporadic aniridia with no other congenital anomalies.[33] One fifth of sporadic aniridia patients may develop WT[34]and one third of patients with sporadic aniridia will have WAGR syndrome.[35] All the cases of WT should be subjected to ocular examination for detection of any associated ocular anomalies.[33]In our case there were no ocular, genitourinary and musculoskelital abnormalities.

The management continues to evolve with two different approaches. Pediatric Oncology Group Protocol (POGP) indicates immediate nephrectomy of affected kidney. However, International Society of Pediatric Oncology (SIOP) advocates the use of neoadjuvant chemotherapy shortly after the clinical or imaging diagnosis.[36] This protocol aims to reduce the rates of surgical complications like tumor rupture and allows selectively performing of laparoscopic surgery.[4] Despite these disparate strategies, the overall survival is comparable.[37] Therefore, a real challenge is not this dilemma but stratification of treatment intensity according to the clinical, histologic and molecular risk factors.[38] In our case,immediate total nephrectomy was performed just after radiological diagnosis followed by 19 cycles of chemotherapy with Vincristine and Actinomycin-D over 18 weeks.

Surgery remains a crucial part of treatment. The indicated surgical procedure is total nephrectomy associated with lymph nodes resection- renal hilum and periaortic, even they do not seem affected macroscopically. In bilateral cases, total nephrectomy must be performed on the side of the larger tumor mass and renal parenchyma should be preserved by enucleation of the tumor mass of smaller size.[4] The current goal is reducing the morbidity associated with chemotherapy, such as cardiomyopathy, renal failure and an increased risk of secondary malignancy.[39]

An overall cure rate of WT is over 85%.[40] However, an increased risk of tumor relapse and/or de novo disease in the contralateral kidney with potential development of renal failure required oncologic follow up at least once a year during childhood.[32]

Annual followup examinations are recommended, including laboratory tests (complete blood count, differential white blood count, liver function tests, renal function tests, urine analysis, CXR), routine physical examination and blood pressure monitoring . Some studies report a high incidence (upto 50%) of unexplained end- stage renal disease occurring approximately 10 yrs after the diagnosis.[41]Our patient was on regular follow up after chemotherapy, initially at 3 months interval for 2 times followed by 6 monthly and will be continued throughout childhood.

IV. Conclusion:

WT is a very well known common renal tumor of pediatric population. But presentation at neonatal period is very rare and uncommon. Here, we report a case of WT presented just after birth with renal mass. Despite well- established literature, studies focused on rare presentation of WT at neonatal period are still needed as well as research aimed on proper prenatal assessment for better knowledge of possibly congenital presentations of WT.

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