

Left Congenital Mesoblastic Nephroma in a Term Male Neonate: A Case Report

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Abstract

Keywords

- Mesoblastic nephroma
- congenital renal tumor
- paraneoplastic syndrome

Congenital mesoblastic nephroma (CMN) is a benign and very rare renal tumor, typically occurring in utero or during infancy. We are reporting a very young case of left sided classical congenital mesoblastic nephroma in a full term, small for gestational age, male neonate; who was detected with left sided flank mass immediately after birth. The patient was managed successfully by total nephrectomy. The diagnosis of CMN-classical type was confirmed on histopathological examination.

Introduction

Neonatal tumors are rare entities comprising nearly 2% of malignancies in childhood. However, congenital mesoblasticnephroma is the most common renal tumor in neonates and in childhood. This tumor was first described in 1967 by Bolande et al.¹ It is diagnosed mostly in the antenatal period with judicious use of ultrasonography or immediately after birth. Nearly 3-10% of renal tumors in children are mesoblasticnephroma with approximately 50% of the cases occurring during the neonatal period diagnosed in the first 3 months after birth. Overall, 90% of cases are discovered by the age of 1 year and almost none occurs after the age of 3 years.² Although it is considered to be benign it can show aggressive behavior leading to catastrophic complications.^(3,4) since it is very similar to other pediatric renal neoplasms; its diagnosis is challenging to both the pediatric surgeons and histopathologists. Also, some CMNs are known to be associated with polyhyrammios, prematurity and paraneoplastic syndromes including hypertension and hypercalcemia.

We are reporting a case of full term, small for gestational age, male neonate who presented with left flank mass immediately after birth and was diagnosed with classical type CMN on histopathological examination. However, no association with paraneoplastic syndromes was found in our case.

Case presentation

A full term (38 weeks), small for gestational age, 1.8 kg male baby was born to a 26-year-old healthy woman (G2, P1, A0) through normal vaginal delivery. The pregnancy was unbooked and the mother had no clinical illness during pregnancy. The delivery was performed at a civil hospital. Immediately after birth, the baby was detected with left sided flank mass and was referred to a tertiary care centre for further evaluation. The baby was brought to the Accident and Emergency Department. A detailed clinical history revealed that no ultrasound had been done during pregnancy and history of exposure to teratogens was also insignificant. General examination revealed heart rate of 152/min, blood pressure of 52/30mmHg and respiratory rate of 68/min with mild subcostal

retraction. On abdominal examination, a mass was palpable in the left flank occupying almost the whole of the left lumbar region. The rest of the examination was normal. Serum creatinine, blood urea nitrogen and serum electrolytes including serum calcium were all normal.

The child was further investigated with abdominal ultrasonography which revealed a well- defined 45x35 mm hyper echoic lesion in the left lumbar region. The mass showed internal vascularity. However, there was no calcification, no vessel encasement and the mass was not crossing the midline. CT scan showed 41x32x43 mm sized heterogeneously enhancing mass lesion in the left kidney without calcification (Figure 1). Probable diagnosis of Wilm's tumor was considered.

passing lower edge of the sternoclavicular joint and following the lower edge of the clavicle which helped to remove the tumor completely with optimal control of noble elements. The evolution was good after two years of follow-up.

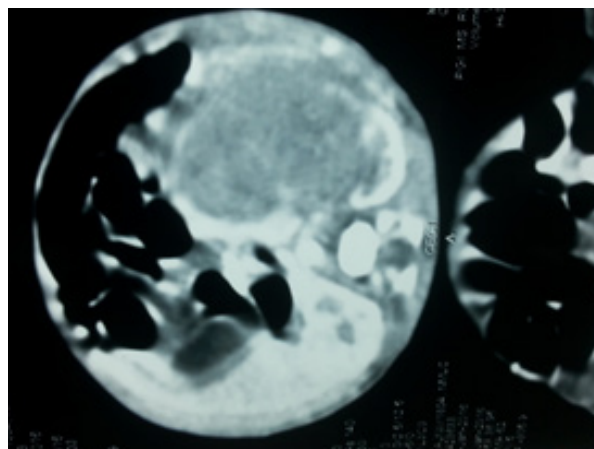


Figure 1: CT scan showing heterogeneously enhancing mass lesion in left kidney

The neonate was transferred to NICU and after cardio-respiratory stabilization and adequate resuscitation, the newborn was taken up for an exploratory laparotomy at the age of 1 week. Through a left supraumbilical transverse incision, abdomen was opened and explored. Liver and the right kidney were found to be normal but there was a solid mass lesion in the left kidney measuring approximately 3.5×3.5×5 cm occupying almost the whole kidney. Left total nephrectomy was performed after ligating the renal pedicle **Figure 2**.



Figure 2: Resected nephrectomy specimen with tumor occupying almost whole of the kidney

After attaining adequate haemostasis, abdomen was closed in layers. The resected specimen was sent for histopathological examination. Gross pathological examination revealed a yellow solid renal tumor with no identifiable necrotic or hemorrhagic areas **Figure 3**.



Figure 3: Photograph showing yellowish renal tumor and whorled cut surface

Microscopic examination revealed a classical type of CMN with no infiltration of the renal pelvis, calyces and the suprarenal gland without any vascular invasion **Figure 4**.

Postoperatively, the patient was kept under observation in NICU. Intravenous fluids and antibiotics were started. Oral feeding was started on the 3rd post operative day. The patient responded well during the admission with all relevant investigations within normal limits. He was discharged 10 days after the operation. He has regular follow ups and is doing well.

Discussion

Congenital mesoblastic nephroma, which is also

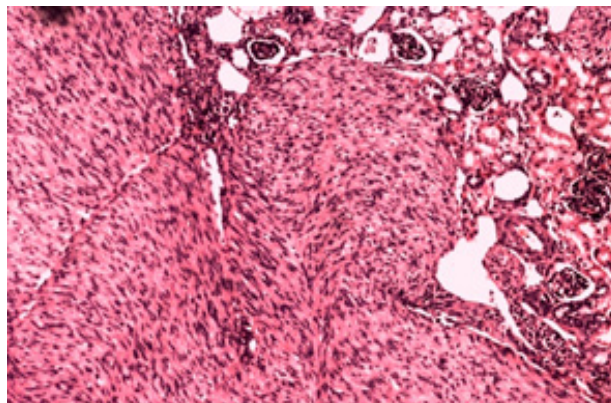


Figure 4: Photomicrograph showing the histopathology of classical congenital mesoblastic nephroma

called fetal renal hamartoma or mesenchymal hamartoma, is considered the most common solid tumor in neonates, corresponding to approximately 3% of all renal neoplasias. It is slightly more common in males and the male-to-female ratio is around 1.5:1.⁵ It tends to occur more commonly in the right kidney. However, in our case, it occurred in the left kidney. An abdominal mass which is palpable and asymptomatic is the most common clinical presentation. Other presentations, though less frequent, are hematuria, proteinuria, polyuria, vomiting, jaundice, dehydration, azotemia and electrolyte disturbances. None of them were seen in our patient.

Histology is an important factor in the diagnosis and predicting the prognosis of CMN.⁶ Based on microscopic appearance, three subtypes are identified namely the classic, cellular and mixed subtypes. These subtypes vary in factors such as: age at presentation, imaging and the biologic behavior. The classic variant is characterized by low cellularity and predominance of matrix with no atypia. It presents earlier, usually before the age of 3 months. Macroscopically the tumor is firm, yellowish gray to tan, with infiltrating borders and a whorled cut surface similar to uterine leiomyoma. Microscopic examination reveals fascicles and whorls of spindle cells with features of secondary mesenchyme and containing vimentin, fibronectin but lacking keratin and laminin. These enlarged cells (myofibroblasts) are seen infiltrating the surrounding renal parenchyma, thus, impairing the growth of renal tubules and glomeruli. However, mitosis is rare

in this variant. It has a better prognosis. On the other hand, the cellular variant is more cellular, being softer with cystic, necrotic and intramural haemorrhagic areas and irregular contours, macroscopically. It has a sarcomatous appearance consisting of tightly packed cells with high nucleus/cytoplasmic ratio, nuclear atypia and frequent mitosis (25-30/10 HPF); thus it tends to have a more aggressive behavior and a worse prognosis. It, however, presents a few months later in life and is more common than classical variant accounting for 42-63% of all cases.⁷ Both variants show negative immunohistochemical expression for epithelial markers and are positive for fibroblastic markers such as vimentin, smooth-cell actin and desmin. But mainly morphological criteria are taken into consideration for diagnosis. Mixed tumors accounts for 10% of the cases and have areas of low and high cellularity with a combination of above features. The histological differential diagnosis mainly includes wilms tumor which usually occurs after the age of 3 years, clear cell sarcoma of the kidney which usually occurs between 2-3 years of age and rhabdoid tumor which occurs between 1-2 years of age.

Image findings in CMN vary in different subtypes since the classic variants are mainly solid, but the cellular variants have a cystic composition. However, the initial investigation is usually ultrasonography. It helps in localizing the tumor and defining the composition of the mass. It usually shows a large, echogenic mass originating from the kidney. Chan et al described a vascular ring sign, an anechoic or hypoechoic vascular ring surrounding the tumor, which can be seen on ultrasonography. It is characteristic of the typical or nonaggressive mesoblastic nephroma.⁸ The diagnosis of mesoblastic nephroma can be achieved in the third trimester by antenatal ultrasonography. CT scan with contrast or magnetic resonance imaging (MRI)

help in the staging of the tumor by showing local extension or distant metastasis if present. They can also help in confirming the origin of the tumor. Turkistani et al reported a case of right mixed congenital mesoblastic nephroma in a 28 weeks premature female with a prenatal history of polyhydramnios and development of hypertension in the first week of life.⁹ Khashu et al reported a case of CMN of right kidney presenting with neonatal hypertension in a premature male neonate.¹⁰ A case of cellular CMN in left kidney was reported in a 2 year old female by Santos et al in 2011. The child underwent nephrectomy but could not survive owing to the aggressive nature and recurrence of tumor.¹¹ We are reporting an uncommonly encountered classical variant of CMN in a term male neonate in left kidney which is less frequently involved.

Total nephrectomy is curative. Complete tumor excision including entire capsule should be considered to prevent subsequent complementary treatment. Nevertheless, an insufficient surgical margin requires a new intervention in order to remove the residual disease. Chemotherapy is occasionally used in cases of residual microscopic disease or in cases of tumor rupture, as well as in rare cases of recurrence or metastases. The overall prognosis of CMN is good with a 5-year survival of 94% and an overall survival of 96% in infants. excision including entire capsule should be considered to prevent subsequent complementary treatment. Nevertheless, an insufficient surgical margin requires a new intervention in order to remove the residual disease. Chemotherapy is occasionally used in cases of residual microscopic disease or in cases of tumor rupture, as well as in rare cases of recurrence or metastases. The overall prognosis of CMN is good with a 5-year survival of 94% and an overall survival of 96% in infants.

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