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Samet Benli *

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Case Report

# Congenital Mesoblastic Nephroma: A Rare Neoplasm of The Newborn

**Samet Benli [1]*, Ahmet Demez [2], Zeynep Vural [3], Mehmet Ali Kopan [3], Ezgi Tanburoğlu Çeliker [3], Serhat Özkan [4], Nazan Tekin [5], Alara Akdeniz [5]**

[1]Cengiz Gökçek Obstetrıcs And Gynaecology Hospıtal, Neonatology Deparment, Gaziantep, Turkey

[2]Gaziantep City Hospital, Department of Paediatric Surgery, Gaziantep, Turkey

[3]Cengiz Gökçek Obstetrıcs And Gynaecology Hospıtal, Pediatrics Deparment, Gaziantep, Turkey

[4]Gaziantep City Hospital, Neonatology Deparment, Gaziantep, Turkey

[5]Gaziantep City Hospital, Pediatrics Deparment, Gaziantep, Turkey

***Corresponding Author:** Samet Benli, M.D, Cengiz Gökçek Obstetrıcs and Gynaecology Hospıtal, Neonatology Deparment, Gaziantep, Turkey

**Received date: January 21, 2025; Accepted date: February 17, 2025; Published date: June 03, 2025**

**Citation:** Samet Benli, Ahmet Demez, Zeynep Vural, Mehmet A. Kopan, Ezgi T. Çeliker, et al, (2025), Congenital Mesoblastic Nephroma: A Rare Neoplasm of The Newborn, *Clinical Research and Clinical Trials*, 12(5); **DOI:10.31579/2693-4779/255**



**Abstract:**

Congenital mesoblastic nephroma (CMSN) is the most common renal neoplasm in the first trimester of the neonatal period (1). The tumor was first described as renal tumor of infancy by Bolande et al. in 1967 and histologically divided into classical, mixed and cellular subtypes (2). The tumor is usually surrounded by renal capsule and the tumor-renal parenchyma border is irregular. It shows a growth pattern spreading to the renal parenchyma and infiltrating the perirenal adipose tissue (3,4). The tumor has a myomatous appearance and has a rubbery consistency. Classic or typical congenital mesoblastic nephroma (or fibromatous type) (1/3) is usually seen before the first 3 months of life and has a benign course.

**Keywords:** congenital; ventilation; parenchyma

## Introduction

Congenital mesoblastic nephroma (CMSN) is the most common renal neoplasm in the first trimester of the neonatal period (1). The tumor was first described as renal tumor of infancy by Bolande et al. in 1967 and histologically divided into classical, mixed and cellular subtypes (2). The tumor is usually surrounded by renal capsule and the tumor-renal parenchyma border is irregular. It shows a growth pattern spreading to the renal parenchyma and infiltrating the perirenal adipose tissue (3,4). The tumor has a myomatous appearance and has a rubbery consistency. Classic or typical congenital mesoblastic nephroma (or fibromatous type) (1/3) is usually seen before the first 3 months of life and has a benign course. The other type, defined as cellular or atypical congenital mesoblastic nephroma (2/3), seen in older infants and children, can be malignant and is capable of recurrence and metastasis. Mixed forms with a combination of the two types have also been reported (5,6). Patients usually present with a palpable abdominal mass, arterial hypertension, hematuria, polyuria or hypercalcemia (7).

## Case Report

A 29-year-old pregnant woman with no prenatal follow-up was found to have polyhydraminosemia on prenatal ultrasonography and the baby was found to have an abdominal mass and dilated intestinal loops. The patient was born at 39 weeks of gestation at 3490 grams by cesarean delivery. APGAR at the 1st minute of delivery was 7 and APGAR at the 5th minute was 8. The patient who underwent positive pressure ventilation after delivery was admitted to the neonatal unit with the diagnosis of respiratory distress and intraabdominal mass. On physical examination, abdominal distension was present and the traube area was closed as pathologic findings. In addition, there was a 9x5 cm mass to the left of the umbilicus consistent with left kidney localization. Whole abdominal ultrasonography of the patient revealed a 90*48 mm mass lesion in the left renal lobe with a relatively smooth bordered mass lesion with solid and cystic components. Contrast-enhanced abdominal CT was performed for further examination. Contrast-enhanced CT scan was reported as a 9x6 cm space-occupying lesion with cystic and solid components in the left renal lobe (Figure 1).

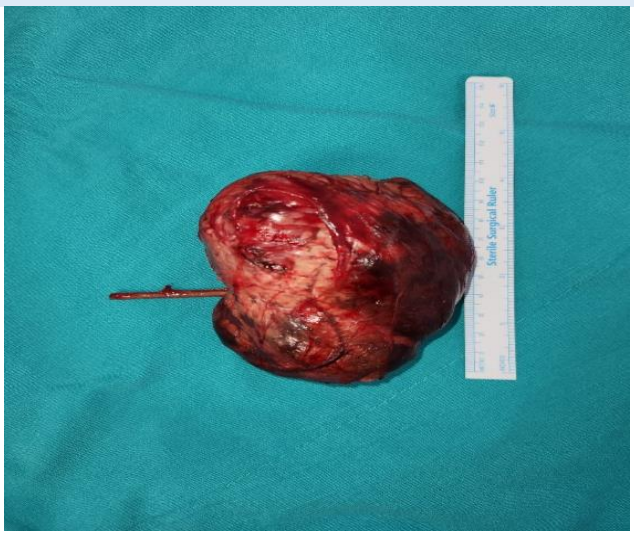


**Image 1:** Macroscopic image of the mass postoperatively

Echocardiography revealed PDA and secundum ASD. Amlodipine 0.5 mg/kg/day was started when blood pressure values were above the 90th percentile on the 3rd postnatal day. The patient was placed under close blood pressure follow-up. During follow-up, the patient's calcium value was 12.1 mg/dL. Calcium value regressed to normal after hydration. Total nephrectomy was performed on postnatal day 18 (Figure 2).

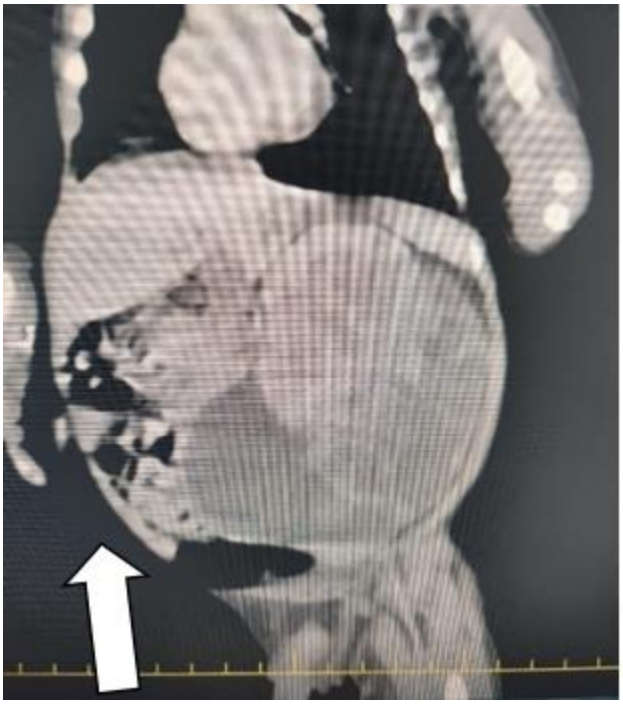

**Image 2:** Abdominal CT, 9x6 cm cystic space-occupying lesion with solid components in the left renal lobe

After total nephrectomy, the patient's blood pressure values were in the normal percentile. The patient was discharged with recommendations on postnatal day 27 with pediatric surgery and pediatric nephrology outpatient follow-up.

## Discussion

CKN accounts for 3-6% of all renal masses in childhood. It is the most common renal tumor of the neonatal period and early infancy. Prenatal diagnosis of fetal tumors is important for maternal and fetal well-being as well as neonatal management. Prenatal ultrasonographic detection of mesoblastic nephroma was first reported by Ehman et al. CKN cases are often recognized as a general renal pathology on antenatal USG and are often accompanied by polyhydramnios, prematurity and neonatal hypertension. Recently, calcium level has been associated with mesoblastic nephroma and hypercalcemia has been proposed as the mechanism underlying polyhydramnios in mesoblastic nephroma (1,8). In our case, hypercalcemia was observed on postnatal day 5 and regressed with hydration. Hypertension was detected on postnatal day 3, antihypertensive treatment was initiated and hypertension returned to normal after the operation.

CGN may present ultrasonographically as a unilateral renal mass, large nodular densities (4 to 8 cm) or diffuse renal enlargement. These tumors are usually solid, but sometimes cystic areas can be seen. In contrast to Wilms' tumor, CPC has a well-demarcated capsule. It may show abnormalities in systems not associated with the BMN tumor. These abnormalities may be associated with neuroblastoma, central nervous system disorders, genitourinary problems, gastrointestinal problems and limb abnormalities. There was no additional pathology in our case. Radical resection of the tumor

or nephrectomy is the treatment of choice. However, local recurrence or distant metastasis due to inadequate resection may occur in a small percentage of patients with BMN. Recurrence or distant metastasis is seen in cellular (atypical) WMNC. While there is a familial predisposition in Wilms' tumor, familial predisposition has not been detected in BMN and this condition is not associated with abnormalities in other chromosomes (6,9,10).

Due to the benign nature of CPC, term or near-term term termination of pregnancy is recommended (1).Our patient was a migrant pregnancy without follow-up and her first examination was performed prenatally at 39 weeks of gestation. Total nephrectomy was performed on neonatal day 18 and the postoperative period was uneventful. Antenatal evaluation of congenital mesoblastic nephroma allows prenatal diagnosis and postnatal management planning to prevent complications (prematurity, developing myocardial decompensation and fetal hydrops). According to the literature, polyhydramnios was observed in approximately 40% of cases of CGM and acute fetal distress occurred in 25% of CGM fetuses (1,11). In our case, there was a history of polyhydramnios on antenatal ultrasonography.

In conclusion, CPC is a tumor with a very good prognosis today with the development of both prenatal and postnatal facilities and the possibility of early presentation to hospital with complications in early infancy. Surgical resection is sufficient for complete cure.

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**DOI:10.31579/2693-4779/255**