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CASE REPORT

Intra-Abdominal Mass in a Newborn: Congenital Mesoblastic Nephroma

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Abstract

Congenital mesoblastic nephroma (CMN) is the most common renal neoplasm seen in infants in the first 3 months of life. It constitutes 3-10% of all childhood renal tumors1. There are three histological types of CMN: classical, cellular, and mixed3. CMN cases are treated with radical surgery (nephroureterectomy) and it has a good prognosis. A 2-day-old female case was presented, with a diagnosis of congenital mesoblastic nephroma who has been referred to us with prenatal intra-abdominal mass.

Keywords: Congenital mesoblastic nephroma; kidney; pediatric neoplasm.

Congenital mesoblastic nephroma (CMN) is the most common renal neoplasm seen in infants in the first 3 months of life. It constitutes 3-10% of all childhood renal tumors[1]. This solid tumor, first described by Bolande et al. in 1967, contains immature mesenchymal cells and a normal renal component[2]. There are three histological types of CMN: classical, cellular, and mixed[3]. The classic typical form (fibromatous) is usually seen in infants under the first 3 months of age and has a benign course. The cellular or atypical form is seen in older infants and children and may have a malignant course. More than half of the cases are of the cellular type. In the differential diagnosis, especially Wilms' tumor, infantile polycystic kidney, multicystic dysplastic kidney, hydronephrosis, focal renal dysplasia, malignant nephroblastoma and diffuse nephroblastomatosis should be considered. CMN cases are treated with radical surgery (nephroureterectomy) and the prognosis is quite good. In this article, we aimed to present a patient diagnosed with CMN, who was referred to us with a prenatal intra-abdominal mass.

Case Report

A 76*82 mm mass in the left kidney was detected in the prenatal ultrasonography of a 2-day old female patient who was born by cesarean section at 37+6 weeks from a 30-year-old mother, with an Apgar score of 6-7/10, and a birth weight of 3240 g. Double marker test and triple antenatal screening tests were negative. Fetal MRI revealed a 60*52 mm mass originating from the left kidney (Fig. 1).

On physical examination of the patient, the abdomen was tense, and a prominent palpable mass was found in the left quadrant. In the biochemical tests performed when he was three days old, hemogram was normal, urea was 12.84 mg/dL, and creatinine was 0.58 mg/dL. High erythrocytes were detected in the complete urinalysis; the urine culture was sterile.

Alpha-fetoprotein was 40582.65 ng/mL (<9 ng/mL), ferritin was 281.38 ng/mL (8.7-71.6 ng/mL), carcinoembryonic antigen was 1.66 ng/mL (<3 ng/mL), vanillylmandelic acid in spot urine test was 10.9 mcg/mg (5.9-37

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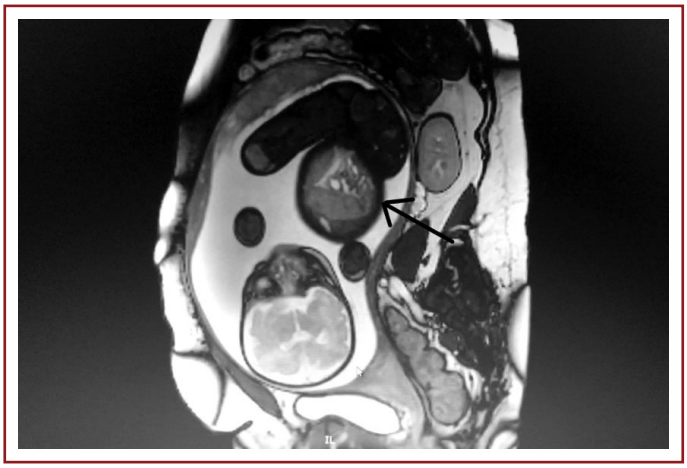

Figure 1. Fetal MRI, 60*52 mm mass appearance in left kidney (Area indicated by arrow: mass in fetal left kidney).

mcg/mg) and neuron-specific enolase was 39 mcg/L (0-16.5 mcg/L).

Echocardiography showed bicuspid aortic valve, atrial insufficiency (minimal) and patent foramen ovale. Amlodipine 0.5 mg/kg/day was started when blood pressure values remained above the 90th percentile on the postnatal 5th day of follow-up. The patient was taken under close blood pressure monitoring. In the abdominal ultrasound sonography performed at the age of 4 days, the mass lesion measuring approximately 85x75 mm, completely infiltrating the kidney on the left, was extending slightly to the right of the midline and arching the aorta to the right. In the abdominal tomography, there was a solid mass lesion of approximately 77x65 mm in size, containing low densities, which may belong to cystic degenerate areas or focal caliectasis, extending to the pelvis on the left, continuing to the right of the midline and arching the aorta.

No filling defect compatible with thrombosis was detected in the left renal vein and inferior vena cava. Several rounded lymphadenopathies were noted in the paraaortic and aortocaval distances, the largest reaching 7.5x7 mm in size (Fig. 2).

No feature was detected in chest computed tomography. The patient was consulted with pediatric hematology-oncology, pediatric radiology, and pediatric nephrology clinics. The patient was operated when she was 11 days old and the mass was totally excised; paraaortic lymph node biopsy was performed (Fig. 3).

Histopathologically, it was reported as classical type CMN with mild atypia and moderate cellularity, no necrosis, no involvement of Gerota's fascia, renal sinus, renal vein and renal capsule.

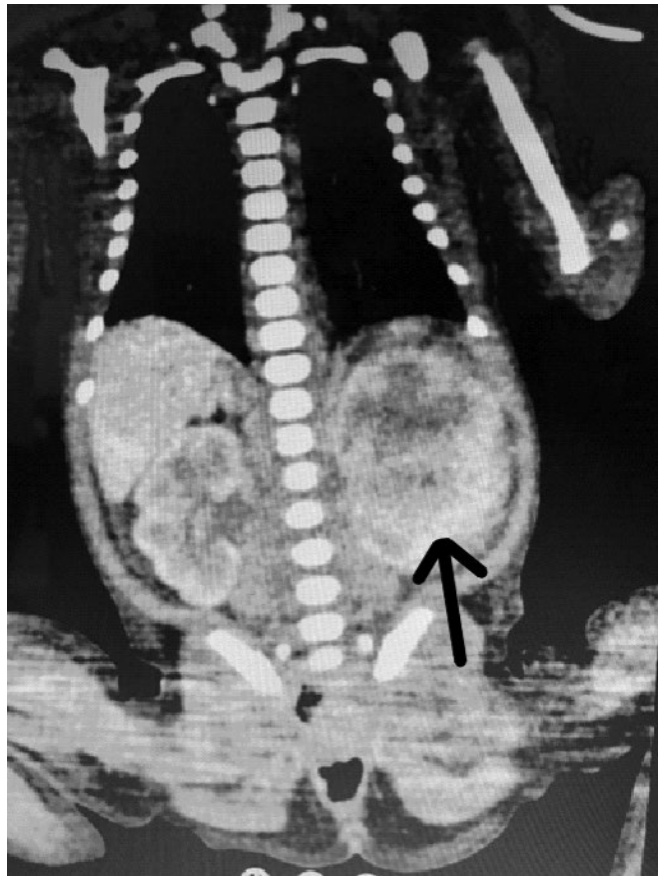

Figure 2. Abdominal CT, 77x65 mm, solid mass appearance with low densities, which may belong to cystic degenerate areas or focal caliectasis.

Right kidney and kidney function tests were found to be normal in the postoperative 24-month follow-up.

Discussion

CMN is a rare childhood renal tumor. It is usually detected by prenatal ultrasonography[4,5]. CMN most commonly presents as an asymptomatic abdominal mass. It can also cause clinical findings with hypercalcemia, hypertension, hematuria and polyuria[6,7]. A possible diagnosis can be made by imaging studies, but histological study is required for definitive diagnosis.

Bayındır et al.[8] found hypertension in 70% of patients. They stated that the high level of renin secreted as a result of trapped glomeruli in the tumor is effective in the formation of hypertension[7,9,10]. The difference in blood pressure measured from the lower and upper extremities was thought to be due to the compression effect of the mass on the aorta.

In our patient, on the postnatal 5th day, the blood pressure values were above the 90th percentile, and the renin aldos-

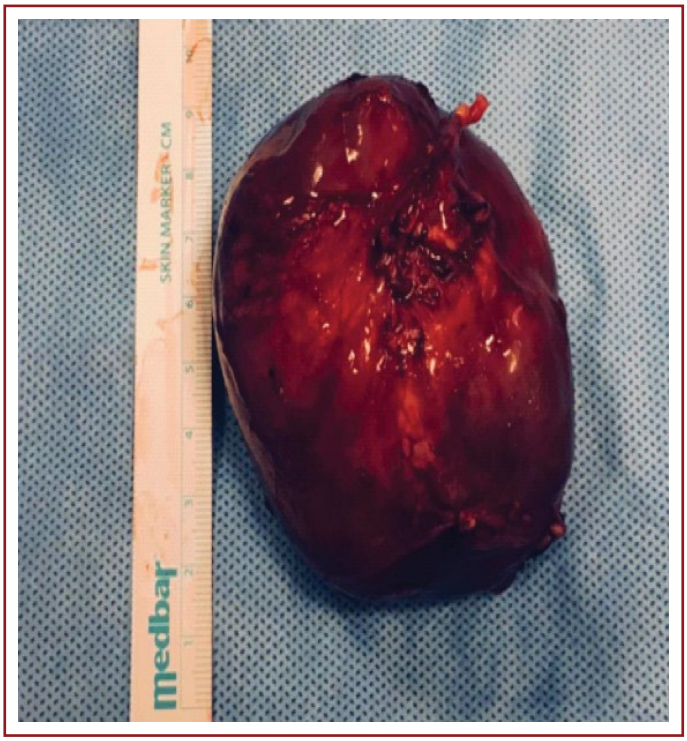

Figure 3. Total excision of the mass.

terone level was measured as normal at 648.3 pg/mL (50-1750 pg/mL). Thereupon, amlodipine 0.5 mg/kg/day was started and close blood pressure monitoring was started. The high blood pressure detected in the patient was attributed to the compression of the aorta by the left renal mass. Amlodipine treatment was discontinued when blood pressure values were found to be normal in the postoperative period. In the patient's follow-up, blood biochemistry values, complete urinalysis and urine culture results were found to be normal.

Hypercalcemia is seen in 1.2%-12% of patients with CMN[11,12]. Hypercalcemia has been associated with the result of the production of parathyroid hormone, parathyroid-related peptide and prostaglandin by tumor cells[11]. In our patient, serum calcium level was found to be within the normal range, at 9.4 mg/dL (7.6-10.4 mg/dL).

Polyuria and hematuria have been reported in some patients with CMN[6,11]. In our case, high erythrocytes were detected in the first postnatal complete urinalysis. No macroscopic or microscopic hematuria was observed in the subsequent examinations. Polyuria was not detected in this case.

CMN is histopathologically divided into subgroups according to the mitosis rate as classical, cellular and mixed[13]. The classical type (24%) represents the presence of a benign mass, while the cellular type (66%) supports aggressive pathology. There is also a mixed type (10%) with characteristics of both types[14,10].

The classic type is usually seen in infants younger than 3 months of age and is characterized by predominantly solid, unencapsulated, fusiform spindle cells, rare mitosis and absence of necrosis[12,15,16]. Histologically, it consists of fibroblastic cells, thin conical nuclei, pink cytoplasm, low mitotic activity, and abundant collagen deposition. In classical types, surgical removal of the mass is curative, and the potential for recurrence and malignancy is low[15,17].

The cellular type has larger areas of bleeding with cystic and necrotic components. The tumor shows high mitotic activity[18]. It has a sarcomatous appearance with oval or fusiform spindle cells and reduced cytoplasm. There is usually perinephric fat and connective tissue invasion. Based on these pathological findings, the cellular type of CMN tends to be more aggressive, and patients with local recurrence or metastasis have been reported[12,13,17]. There is a possibility of misdiagnosis due to the similarity of the cellular type with other common kidney tumors. Therefore, the authors consider this to be an underreported childhood neoplasm[19].

Radical nephroureterectomy is the mainstay of treatment in CMN. Adjuvant chemotherapy is recommended for incomplete removal during surgery, tumor rupture, local recurrences, and distant metastases (cellular and mixed type), which constitute only 5-10% of cases[18,20–22]. Most recurrences occur within one year of resection. Therefore, regular clinical follow-up and abdominal ultrasonography scans are important in follow-up.

The histopathological diagnosis of our case was classical type, which is characterized by moderate cellularity, mild atypia, and absence of necrosis. The important finding in this case was the presence of hypertension in the preoperative period. However, symptoms such as hypercalcemia, hematuria, and polyuria were absent.

This case highlights the importance of evaluating CMN in cases with neonatal hypertension. Significant hypertension in the upper extremities and acute decrease in postoperative blood pressure suggested aortic compression of the mass as a mechanism of hypertension in CMN. Surgical resection is the most common treatment modality to alleviate the paraneoplastic symptoms of the tumor, but it should be noted that the histopathological features of the tumor are the main determinants of further follow-up and treatment of CMN.

Informed Consent: Written informed consent was obtained from the patients' family for the publication of the case report and the accompanying images.

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