Case Report

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A massive primary congenital mesoblastic nephroma was successfully managed through open total nephrectomy: a case report

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Background: Congenital mesoblastic nephroma is the most common renal tumor in children under the age of 6 months, comprising 3–10% of all kidney tumors in children. It is a rare and mostly benign tumor. It divides into cellular, classic, and mixed subtypes. It is typically detected in the third trimester of pregnancy using ultrasonography and magnetic resonance imaging. The best treatment is surgically by completely removing the tumor.

Case presentation: We reported a case of a one-day-old female who was born at 31 weeks gestation weighing 1670 g. Preterm labor was due to polyhydramnios, which was diagnosed predelivery. A large mass was detected in the left hypochondrium using computerized tomography, total nephrectomy was performed, and the histopathological examination of the specimen confirmed the diagnosis of congenital mesoblastic nephroma.

Clinical discussion and conclusion: Early polyhydramnios could be the most significant sign of renal tumors in infants, especially congenital mesoblastic nephroma.

Keywords: case report, CMN, congenital mesoblastic nephroma, kidney tumor, surgery, ultrasound

Introduction

Congenital renal tumors are uncommon neoplasms, including congenital mesoblastic nephroma (CMN)[1]. It is a rare pediatric kidney tumor that represents 3–10% of all pediatric renal neoplasms and peaks in incidence during the first 3 postnatal months[2]. The first definitive sign that a renal lesion is present is polyhydramnios, which causes preterm delivery[3]. Ultrasonography (US) can only identify 11–14% of cases[4]. This tumor may lead to major complications such as polyhydramnios, neonatal hypertension, preterm labor, and hydrops fetalis[2]. The treatment of choice is surgical resection[1]. Herein, we report a CMN case that was successfully managed via total nephrectomy.

HIGHLIGHTS

- Congenital mesoblastic nephroma is a rare tumor that could be detected in the third trimester.
- Total nephrectomy is the ideal treatment for the tumor.
- Polyhydramnios is the most common complication that can lead to preterm labor.

Case presentation

A one-day-old female was born at 31 weeks gestation, weighing 1670 g. APGAR score was within the normal range. Preterm labor was due to polyhydramnios, which was diagnosed predelivery. However, the patient did not need any respiratory support. On examination, abdominal distention was observed, which was particularly more pronounced on the left side. US of the abdomen and pelvis showed a large mass extending from the left hypochondrium (LH) to the pelvis, occupying most of the abdomen's left half and appearing parallel to the left kidney's upper pole. The mass was slightly hyperechoic and heterogeneously contained multiple fluid-filled centers. It showed frank perfusion via Doppler. Its upper part consists of several thin-walled cystic formations, some of which showed clear liquid content, and some showed thick liquid content. The mass measures ~6.2, 4.8, and 5 cm. A computerized tomography (CT) scan of the abdomen and pelvis with contrast revealed a significantly perfused large mass that filled the LH and the left iliac fossa. The mass was pressing on the intestines, stomach, and bladder

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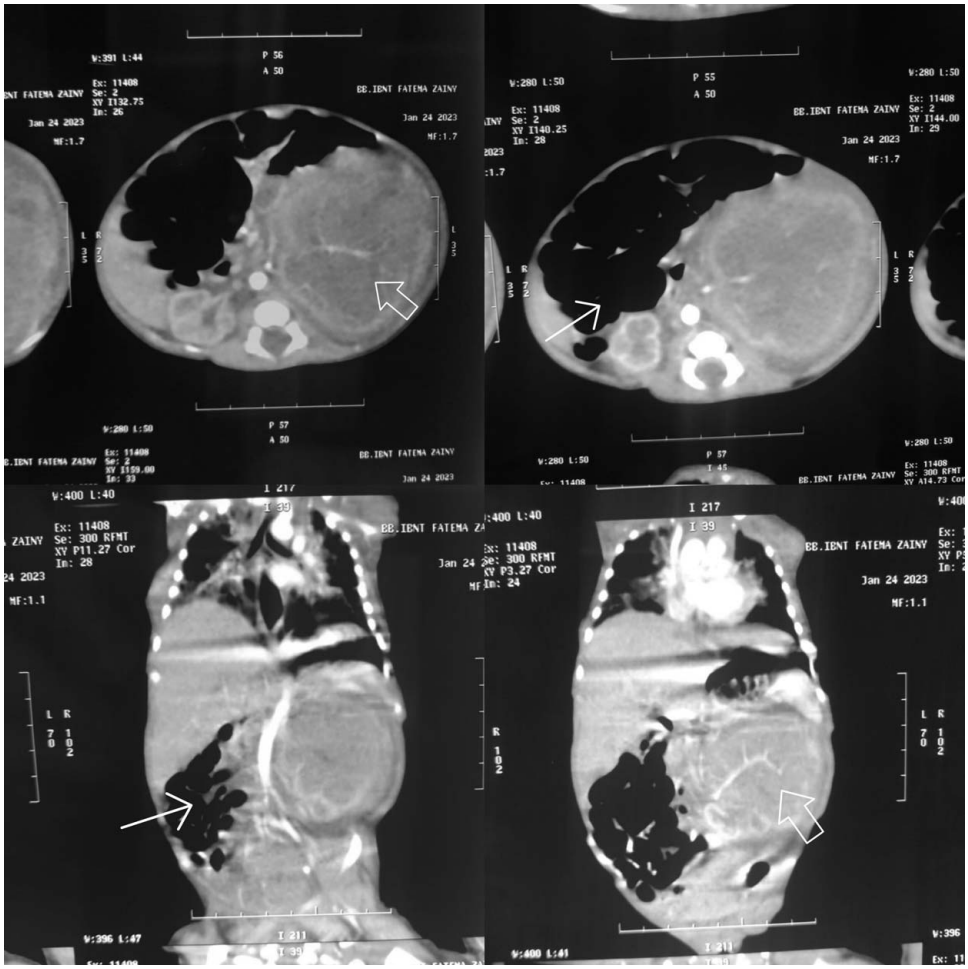

Figure 1. Computed tomography scan revealed a mass lesion (wide arrow) protruding from the upper part of the left kidney's parenchyma showing a broad, hypodensified center with a strongly reinforced periphery and numerous vascular-arterial extensions within it. In its upper anterior part, it contains several cystic formations and strongly pushes the left kidney, intestinal loops, and adjacent anatomical elements (narrow arrow). It appears isolated from the abdominal aorta and measures about (5 × 4.8 × 6.4 cm). A small amount of free fluid is seen in the pelvis.

due to its large size. As a result, the kidney's structure was abnormal, the pancreas was indistinguishable, and the entire intestine shifted to the abdomen's right side (Fig. 1). On the third day of life, a total nephrectomy was performed through an abdominal approach (Video 1, Supplemental Digital Content 1, http://links.lww.com/MS9/A280), due to bile abundance secretions in the nasogastric tube (NGT). The colonic vessels and lower mesogastric artery were dissected and completely isolated to the medial side. Adhesions between the mass, spleen, and pancreas were dissected properly without any damage (Fig. 2). Adrenal gland was completely resected. The specimen showed proliferated spindle cells, fibrosclerotic tissue, hyaline deposit, proliferated blood vessels, local fat tissue at the periphery, and no evidence of atypicality, confirming the diagnosis of CMN (Fig. 3). After 2 weeks, the infant was discharged from the hospital in good general condition and has since been undergoing appropriate follow-up without experiencing any complications.

Discussion

Bolande *et al.* first referred to CMN as a renal benign tumor of infancy in 1967[1]. The renal origin accounts for about two-thirds of infantile or fetal abdominal tumors. The majority of tumors are benign and connected to multicystic dysplasia and hydronephrosis. Until the age of 6 months, CMN is the most prevalent kidney tumor in fetuses and newborns[2]. A significant increase in tumor size occurred in the third trimester when practically all fetal CMN cases were discovered. According to epidemiological research, CMN is more likely to affect the right kidney, and males with a male-to-female ratio of roughly 2:1[5,6]. Infants with CMN have a 5-year survival rate of 94% and an overall survival rate of 96%. However, the prognosis in the cellular variant is worse, which has a 5-year survival rate of 85%[7]. Our patient was born at 31 weeks of gestation, weighing 1670 g, and had severe prematurity. The patient had a massive and abundant ischemic mass covering the left kidney's upper pole and extending from the left hypochondrium to the left iliac fossa (Fig. 1).

Polyhydramnios affects 15–36.4% of CMN patients and increases the risk of preterm delivery. Polyhydramnios can be caused by excessive blood perfusion in the kidney and constriction of the surrounding intestine[4,8]. In this case, an excessive amniotic fluid was discovered in the third trimester that led to premature labor. Clinically, CMN presentations are similar to other juvenile kidney neoplasms. The majority of CMN cases after infancy have

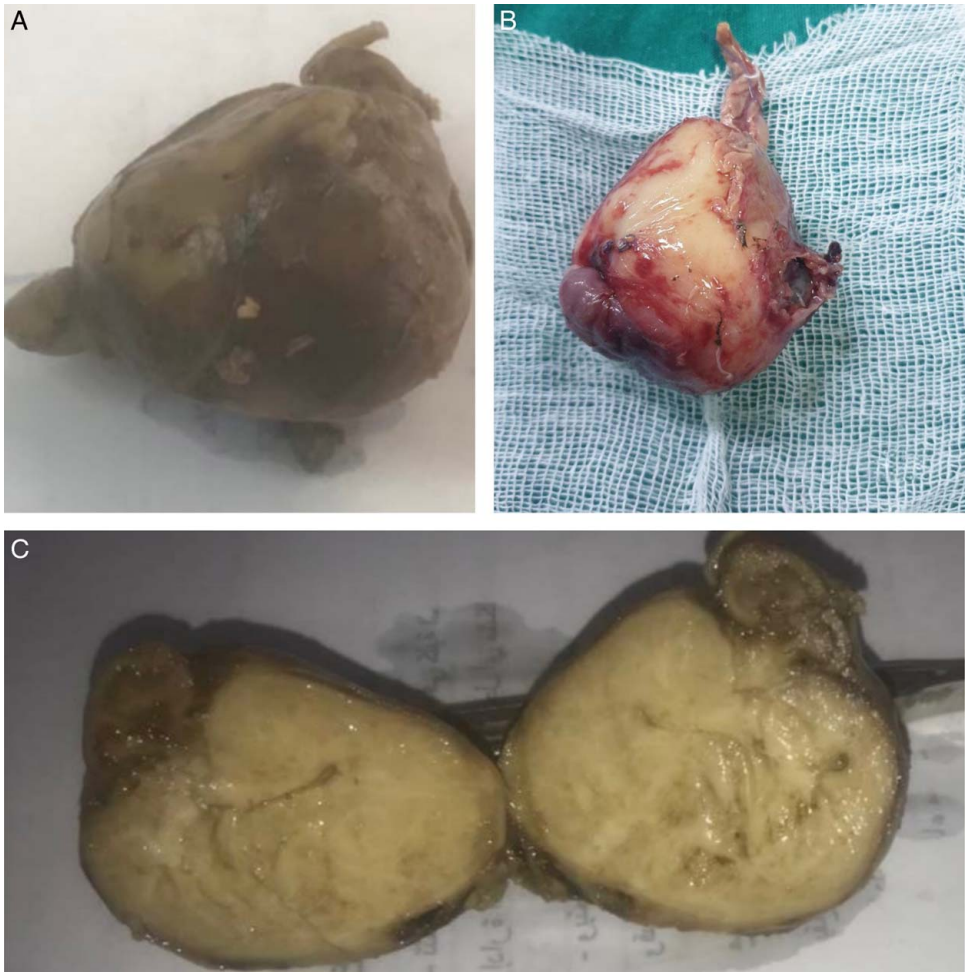

Figure 2. (B) An image showing the mass after resection. (A, C): An image showing the mass during pathological examination.

been discovered incidentally as an abdominal mass and distension; however, other symptoms have included abdominal and flank pain, +\– vomiting, weight loss, loss of appetite, polyuria, hypercalcemia, hypertension, and congestive heart failure[9]. In our case, the patient had a normal Apgar score, and the NGT excessive bile secretions were the major symptom due to tumor-aggressive pressing on the intestine, stomach, and spleen, pushing the bladder toward the pelvis. In addition, there were significant adhesions between the mass and pancreas, spleen, and kidneys. The pancreas was also difficult to distinguish because of the mass size, which caused an intestine shift to the belly's right side.

CMN can be detected during the second and third trimesters. However, most cases are diagnosed as an abdominal mass in the first 6 months of life[9]. In our case, although an elevated level of amniotic fluid was observed 2 months before delivery, and polyhydramnios was diagnosed during the third trimester, the mass was not detected until the first day after birth. Magnetic resonance imaging (MRI) and US are most used for diagnosing CMN. The relationship between the tumor and adjacent structures can be assessed by MRI. On the US, features of CMN might correspond to a certain histological type. For instance, a solid hypoechoic mass with a 'ring sign' which represents an echogenic rim, could indicate classical CMN, whereas the cellular type indicates a renal mass with cystic degeneration, necrosis, and bleeding[5,10]. In our case, US of the abdomen and pelvis revealed a slightly hyperechoic mass, which contains multiple heterogeneous fluid centers that are vascularized on Doppler. CT with contrast could reveal a mass with margins of less enhancement of contrast than normal parenchyma, but there is no specific radiographic finding generally[9]. In this instance, CT with contrast of the abdomen and pelvis demonstrated a highly perfused and enlarged mass with cystic degeneration on the left kidney's upper pole that compresses the intestines, stomach, and bladder. It is often difficult to differentiate between Wilms tumor and CMN when necrosis and hemorrhage are present. Although the presence of a well-defined capsule suggests Wilms tumor, the absolute distinction is only confirmed after microscopic examination[5,9]. Histopathology, CMN, is divided into three types: classic, cellular, and mixed type. The classical type is characterized by spindle-shaped cells arranged in fascicles, low mitotic activity, and no necrosis. In contrast, cellular variants usually show spindle cells with scant cytoplasm and high mitotic activity[11]. In our case, gross examination revealed a solid yellow-gray mass occupying the upper lobe. Histological study showed proliferated spindle cells with fibrosclerotic tissue and hyaline deposits. Increased vascular proliferation was also observed, while no signs of atypia or necrosis were present. These findings are suggestive of the classical type. Stages I/II CMN

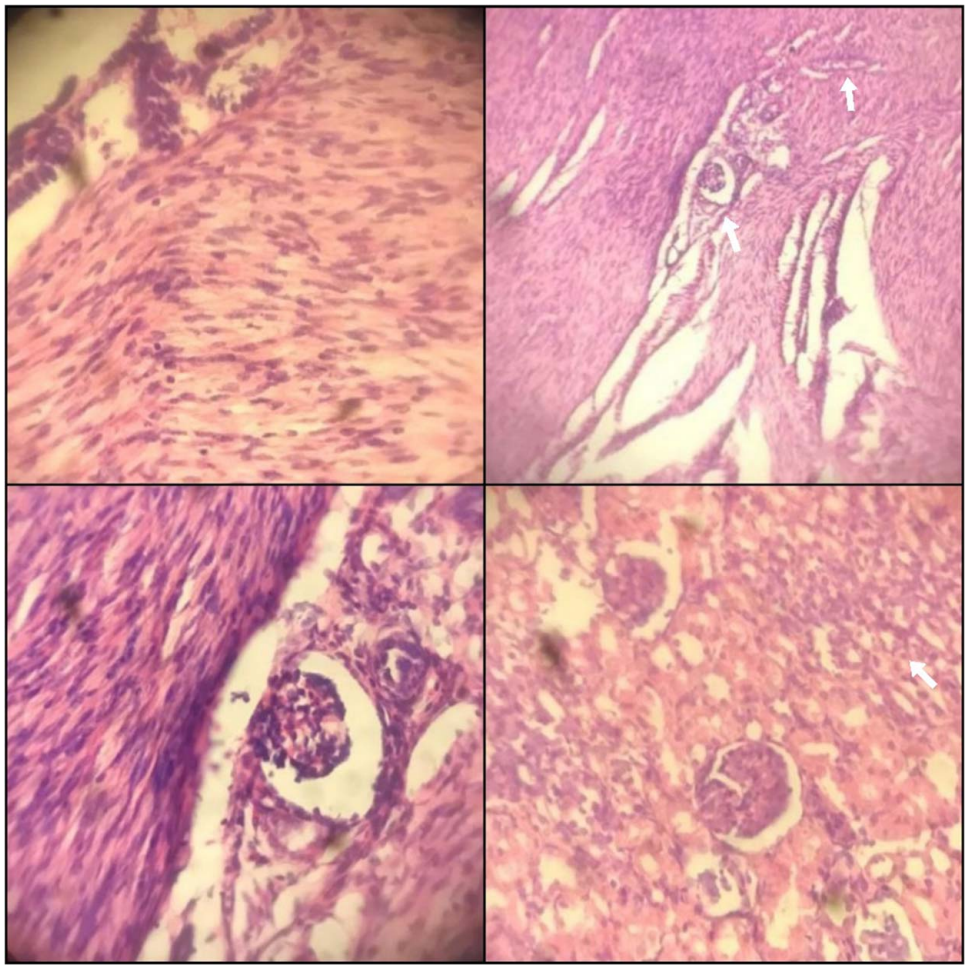

Figure 3. Photomicrograph hematoxylin and eosin (H&E stain) showed proliferated spindle cells, fibrosclerotic tissue and hyaline deposit, proliferated blood vessels, local fat tissue at the periphery, and no evidence of atypicality.

indicate negative surgical margins, while positive margins are suggestive of stage III. Stages IV and V refer to metastasis and bilateral renal involvement, respectively[8]. Complete surgical resection is performed in the first week to treat CMN patients[9]. Patients older than 3 months at diagnosis with stage III, cellular or mixed subtype, or incomplete resection are more likely to experience recurrence and metastases[3,10,11]. Adjuvant chemotherapy is advised for such conditions[12]. Postoperative complications include bowel occlusion and enterocutaneous fistula. The prognosis for CMN is good. However, imaging follow-up for 2 years is recommended post-operation[13]. In our case, a total nephrectomy was performed and the nearby tissues were separated from the mass without chemotherapy. She was discharged after 2 weeks, with no complications on subsequent follow-up.

Conclusion

Polyhydramnios is a common finding in infants with renal malignancies, especially CMN. Although some radiographic characteristics may be useful in diagnosis, pathological examination provides the definitive diagnosis.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the parents of the patient for publishing this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

R.S.: design of the study, data collection, data interpretation and analysis, drafting, critical revision, and approval of the final manuscript; M.M.: design of the study, data collection, data interpretation and analysis, critical revision, drafting, and approval of the final manuscript; Y.A.: data interpretation and analysis, critical revision, drafting, and approval of the final

manuscript; B.A., R.M., and A.A.: drafting, critical revision, and approval of the final manuscript; R.A.: the co-supervisor, drafting, critical revision, and approval of the final manuscript; A.B.: the supervisor, patient care, drafting, critical revision, and approval of the final manuscript.

Conflicts of interest disclosure

There are no conflicts of interest.

Research registration unique identifying number (UIN)

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Guarantor

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Data availability statement

Not applicable. All data (of the patient) generated during this study are included in this published article and its supplementary information files.

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