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A Case Report on Congenital Mesoblastic Nephroma

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Authors' contributions

This work was carried out in collaboration among all authors. Author RAH contributed to the conceptualization and design of the study, performed the literature review, and drafted the initial manuscript and played a key role in the surgical management of the patient and in gathering clinical data. Author ISI assisted in the clinical management of the patient, including imaging studies and histopathological evaluations, provided critical revisions to the manuscript for important intellectual content and ensured the accuracy of clinical information. Author MIAA conducted a comprehensive review of the literature regarding congenital mesoblastic nephroma and contributed to the discussion section and ensured that all relevant findings were included in the manuscript and assisted with final revisions for publication standards. All authors read and approved the final manuscript and agree to be accountable for all aspects of the work, ensuring that any questions related to the accuracy or integrity of the work are appropriately investigated and resolved.

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Case Report

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ABSTRACT

Aims: This study aims to provide a comprehensive review of the epidemiology, genetic underpinnings, clinical presentation, and management strategies of congenital mesoblastic nephroma (CMN), with a focus on the diagnostic and therapeutic challenges that distinguish it from other neonatal renal tumors such as Wilms' tumor. By presenting a unique case of a neonate diagnosed with CMN and undergoing tumor resection, we seek to highlight the complexities of perinatal care in such cases and the potential for life-threatening complications despite early intervention. Furthermore, the study underscores the importance of early and accurate differentiation between CMN subtypes, as well as the role of histopathology in guiding treatment approaches.

Presentation of Case: A female neonate was born at 34+2 weeks gestation, presenting with a retroperitoneal mass on the left kidney, identified prenatally via ultrasound. Delivered by cesarean section due to polyhydramnios and anemia, a palpable intraabdominal mass was noted during examination. Imaging confirmed a heterogeneous solid lesion suggestive of nephroblastoma. The patient underwent nephroureterectomy, with histopathology confirming congenital mesoblastic nephroma.

Despite successful resection, the patient died at 9 days of age from refractory catecholamine septic shock, with complications including neonatal sepsis, gastrointestinal bleeding, and femoral artery thrombosis.

Discussion: Congenital mesoblastic nephroma is the most common renal tumor in neonates, often misdiagnosed as Wilms' tumor. It typically presents before 2 months of age and is associated with translocation (12;15)(p13;q25). The prognosis for classical CMN is excellent, while the cellular variant is more aggressive. Surgical excision is the primary treatment, but the neonate's age necessitates careful management to mitigate risks. In this case, multiple complications contributed to the poor outcome despite timely intervention.

Conclusion: CMN is a rare renal tumor in neonates, requiring accurate diagnosis through histopathology. While prognosis is generally favorable, complications from prematurity can adversely affect survival.

Keywords: Congenital mesoblastic nephroma; Bolande's tumour; nephroureterectomy; neonatal renal tumor.

1. INTRODUCTION

1.1 Mesoblastic Nephroma

Congenital mesoblastic nephroma (CMN) or Bolande's tumour is a mesenchymal tumour of the neonates. It was first described in 1967, distinguishing it from other renal tumour such as Wilm's tumour. Other names of mesoblastic nephroma includes 'fetal renal hamartoma' or 'leiomyomatous renal hamartoma' due to similar whorled pattern like uterine leiomyoma [1,2]. Based on epidemiology, congenital mesoblastic nephroma is considered rare among the population with only about 3% cases found among pediatric renal tumour [3]. However, it is the most common kidney tumour in neonatal period especially in the first 2 months of life. This was one of the main difference between Bolande's tumour and Wilm's tumour, with Wilm's tumour is more commonly found in the age of 2 to 3 years old [1,2]. Gender predisposition of congenital mesoblastic nephroma is also different from Wilm's tumour, as congenital mesoblastic nephroma were predominantly found in male compared to female [1,4].

Genetic study of congenital mesoblastic nephroma is usually associated with translocation of (12;15) (p13;q25) which leads to *ETV6* and *NTRK3* fusion, and a trisomy 11 [3–5]. These genetic anomalies were found in mixed/cellular type CMN. Other genetic anomaly related to CMN such as trisomy 8, 17, 20, 7, 10, 18 and 9 had been reported. Findings of t(12;15)(p13;q25) could give a better prognostic factor due to low recurrence after surgery [1,3]. CMN could be suspected as early as the 3rd trimester of pre natal period through routine ultrasound checkups in pregnancy [2,4]. Classification of CMN based on histology are divided into classic, cellular and mixed type [6,7]. Classical CMN can infiltrate renal parenchyma without causing any hemorrhage or necrosis. Cellular CMN has a high mitotic activity and

invasive tendency. Mixed type has a combination of both classical and cellular features [2].

Overall prognosis of CMN is excellent with classical CMN has better prognosis compared to other types. Cellular CMN may present less favorable outcome due to its aggressive nature. Surgical management is the mainstay therapy for CMN especially in early stages. However, due to its predominant population in neonates, surgery must be considered carefully to avoid potential morbidity from anesthetic or intraoperative complication [1,5]. Neoadjuvant chemotherapy could be considered if there are concerns regarding safety in surgical management [3]. In this case report, we present a case of CMN in female neonate discovered with prematurity and abdominal mass who undergone a nephroureterectomy after a series of renal tumour imaging [6,7].

2. PRESENTATION OF CASE

A female neonate was born with a retroperitoneal mass on her left kidney. The mother was diagnosed with G3P10011 gestational age of 34+2 weeks, shoulder presentation, Premature Rupture of Membrane (PROM) 8 hours, suspicion of fetal nephroblastoma (S), polyhidramnion, anemia (Hb 8,2). The mode of delivery was caesarean section. From physical examination, there is a palpable intraabdominal mass in left hypochondriac and left lumbar with poorly defined border. The intraabdominal mass was suspected for nephroblastoma with differential diagnosis of splenomegaly.

The babygram showed a non homogenous opacity with poorly defined border, well defined margin in left lumbal region that is projected at VL1-VL4 at left side with impression of pressing the intestinal system to right side with suspicion of splenomegaly and differential diagnosis of a mass.

The Multislice Computerized Tomography (MSCT) showed that there is a heterogeneous solid lesion with well defined border partial well defined margin occupying the left hypochondriac region, left lumbar, left iliac, to the left umbilical region, with the impression of originating from the left renal parenchyma, superior and medial poles, which on post-contrast appeared to have heterogeneous contrast enhancement, pressing the surrounding intestinal system, pressing the spleen superiorly, attaching to and pressing the abdominal aorta to the right side, the left common iliac artery to the right side, the lesion appears to receive arterial feeding from the left renal artery, which could be a nephroblastoma. There is also multiple irregular lymphadenopathy in both side of inguinal region.

Surgery was conducted in this patient. Laparatomy incision was made on left supraumbilical above the mass. Left retroperitoneal mass with 7x6 cm in size was found during the surgery, and after retroperitoneal incision, left kidney mass was found. Nephroureterectomy was done in this patient. After mass sample retrieved, it was then sent to anatomical pathology for further investigation. Histopathology result showed that the mass is of mesenchymal origin, confirmed as a mesoblastic nephroma.

Patient died at the age of 9 days with direct causes of death from refractory catecolamine septic shock. Indirect causes of death from neonatal septic, gastrointestinal bleeding, thrombosis of femoral artery. The patient also suffers from prematurity, low birth weight, hyaline membrane disease, mesoblastic nephroma, and pneumonia [8-10].

3. DISCUSSION

The patient was initially suspected to have a mass based on an ultrasound examination of the mother’s pregnancy before delivery. After the patient was born via cesarean section, this suspicion was further confirmed by the discovery of a mass in the abdomen, specifically in the left hypochondrium and left lumbar area. Additional examinations, including a babygram and MSCT, confirmed the presence of a renal mass. Following the retrieval of the mass during surgery and subsequent histopathological analysis, it was concluded to be mesoblastic nephroma.The examined tissue measures 7x8x6 cm, has a resilient texture, and adheres to the ureter at the edge, with two fragments and three fragments visible on cross-section. Microscopically, the adrenal tissue shows no tumor, while a mesenchymal tumor is present between the renal parenchyma. The tumor cells exhibit proliferation with a fascicular appearance, characterized by slightly apical nuclei and a spindle shape. Mitosis is absent, and there is no necrosis, with no signs of malignancy detected. The conclusion or diagnosis indicates the kidney contains mesoblastic nephroma.

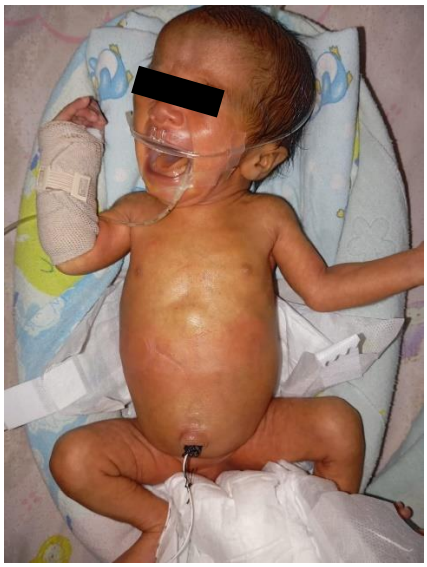

Fig. 1. Clinical presentation of the patient

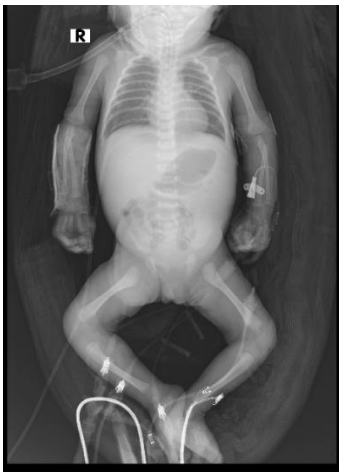

Fig. 2. Babygram of the patient

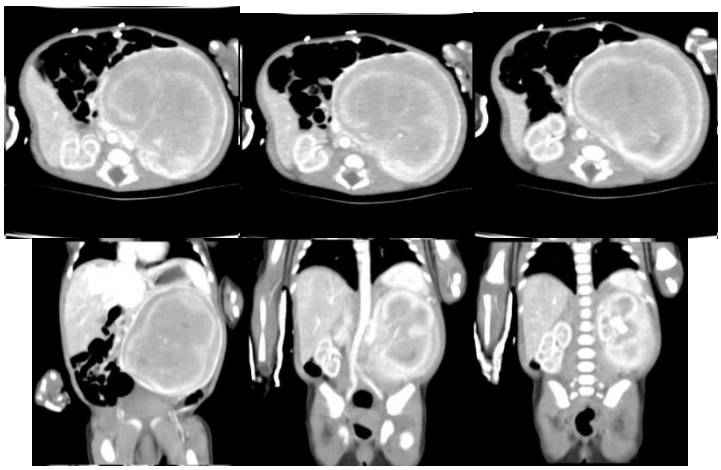

Fig. 3. Abdominal CT scan of the patient

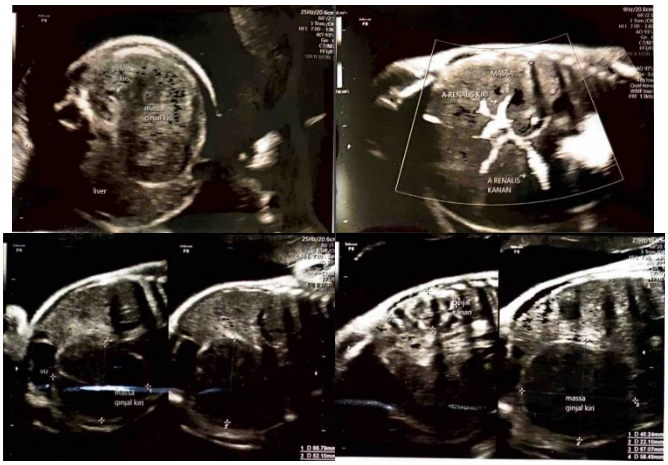

Fig. 4. Antenatal ultrasound of the patient

Mesoblastic nephroma is often accompanied by polyhydramnios (15% - 36.4%), which increases the risk of premature birth. Both of these conditions occurred in this patient. The translocation mentioned is common and shared with infantile fibrosarcoma, a fact that is not described in the study. Prognostically, because the tumor was found at a younger age, the prognosis is generally good. However, the patient's overall prognosis was worsened due to complications of prematurity, such as low birth weight, respiratory distress syndrome, pneumonia, sepsis, gastrointestinal bleeding, and femoral artery thrombosis.

The standard management for patients with mesoblastic nephroma involves surgery, with neoadjuvant chemotherapy as an alternative if there are concerns regarding the procedure. In this case, a nephroureterectomy was performed. During surgery, a 7x6 cm retroperitoneal mass was found, originating from the left kidney. There were no intraoperative complications; however, the patient's condition deteriorated post-surgery, ultimately leading to death.

The complications in this case significantly impacted the patient's outcome, despite early detection and appropriate surgical intervention. Prematurity played a critical role, as the patient was born at 34+2 weeks gestation and faced complications commonly associated with preterm birth. These included low birth weight and respiratory distress syndrome, which increased vulnerability to further issues such as pneumonia and sepsis. Additionally, gastrointestinal bleeding and femoral artery thrombosis further compromised the patient's stability post-surgery. Although congenital mesoblastic nephroma typically carries a favorable prognosis, the combination of these severe complications, particularly septic shock and multi-organ failure, ultimately led to the patient's death. This case highlights the complex interplay between tumor management and neonatal complications, emphasizing the need for vigilant postoperative care, especially in preterm infants [11-14].

4. CONCLUSION

Congenital mesoblastic nephroma is a type of renal cancer most commonly found in neonates. Mesoblastic nephroma can be misdiagnosed with nephroblastoma, so further confirmation by histopathology is needed to differentiate these tumors. Mesoblastic nephroma has an overall excellent prognosis, with classical type better than cellular type. In this case, the patient with mesoblastic nephroma also presented with many other conditions that affect the survivability of this patient.

CONSENT

Informed consent was obtained from the parents of the neonate prior to the surgical procedure and the use of clinical data for this case report. The parents were provided with detailed information regarding the diagnosis, treatment options, and potential outcomes associated with congenital mesoblastic nephroma. They acknowledged their understanding and agreed to the publication of the case details while ensuring the anonymity of the patient. The authors confirm that all ethical considerations have been adhered to in compliance with institutional guidelines and regulations.

ETHICAL APPROVAL

Ethical approval for this case report was obtained from the Institutional Review Board (IRB) at the Medical Faculty, Sebelas Maret University, Surakarta, Indonesia. The study was conducted in accordance with the ethical principles outlined in the Declaration of Helsinki. The authors ensured that all patient information was handled confidentially and that identifying details were omitted to protect the privacy of the patient and family. The IRB reviewed the study protocol and granted approval, recognizing the importance of this case report in contributing to the understanding and management of congenital mesoblastic nephroma in neonates.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

The author(s) hereby declare that no generative AI technologies, including but not limited to Large Language Models (such as ChatGPT, COPILOT), or text-to-image generators, were utilized in the writing or editing process of this manuscript.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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