Case report

CONGENITAL MESOBLASTIC NEPHROMA : A CASE REPORT

ABSTRACT

Aims

This study aims to review the epidemiology, genetic basis, clinical presentation, and management of congenital mesoblastic nephroma (CMN), highlighting its challenges and presenting a case of a neonate undergoing tumor resection.

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.

INTRODUCTION

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. 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. 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. 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.

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.

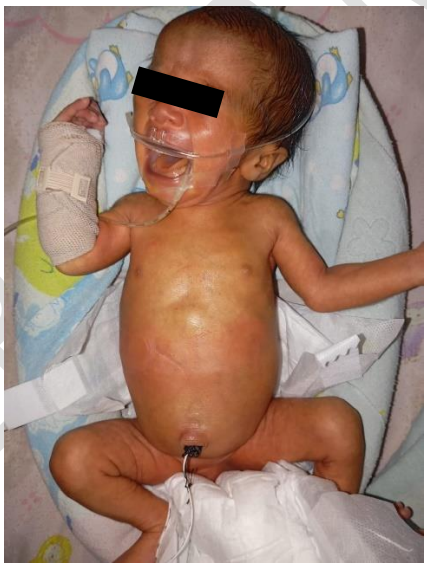

Figure 1. Clinical presentation of the patient

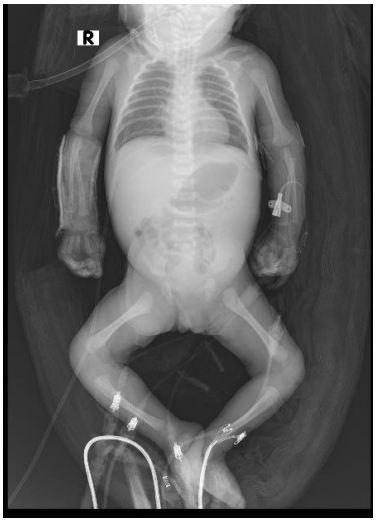

Figure 2. Babygram of the patient

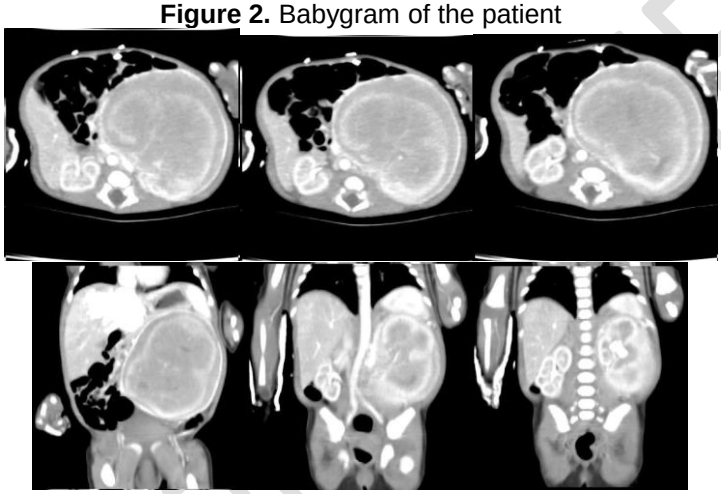

Figure 3. Abdominal CT scan of the patient

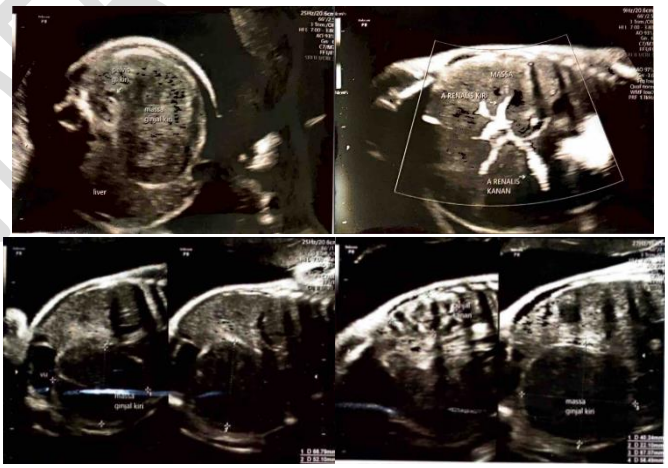

Figure 4. Antenatal ultrasound of the patient

DISCUSSION

The patient was suspected with a mass at first. This was based on an ultrasound examination of the mother's pregnancy before delivery. After the patient was born via cesarean section, the suspicion

was further confirmed by the discovery of a mass in the abdomen, specifically in the left hypochondrium and left lumbar area. Subsequently, additional examinations including a babygram and MSCT confirmed the suspicion of a renal mass. After the mass was retrieved from surgery and analyzed histopathologically, the mass was concluded to be mesoblastic nephroma. This type of tumor is the most common congenital mesenchymal tumor in neonates.[1,3]

Mesoblastic nephroma is often accompanied with polyhydramnios (15% - 36.4%) which increased premature birth risk.[2] Both of these conditions occurred in this patient. Prognostically, because the tumor was found at a younger age, the prognosis is generally good.[1,3] However, the patient's overall prognosis is 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.[2,5] In this case, a nephroureterectomy was performed. During the surgery, a 7x6 cm retroperitoneal mass was found, and after retroperitoneal incision, it was determined to originate from the left kidney. There were no intraoperative complications during the surgery. However, after the procedure, patient's condition deteriorated, and the patient passed away.

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.

COMPETING INTERESTS

The authors declare that there are no competing interests related to this case report and its findings. There has been no financial support or funding received from any organization or entity that could influence the results or interpretation of this study. The authors have no personal or professional relationships that could be perceived as influencing the research outcomes presented in this report. All efforts have been made to ensure transparency and objectivity in the research process and the reporting of findings.

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.

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