CASE REPORT

Congenital Mesoblastic Nephroma at Birth: A Case Report and Detailed Analysis

Hina Yousuf[1*] and Aisha Bakhtiar[1]

[1]*Liaquat National Hospital and Medical College, Karachi, Pakistan*

Abstract

Renal tumours in infants and children, ranging from benign to highly malignant, present a significant medical challenge and constitute approximately 3% of all kidney tumours in neonates. This study focuses on congenital mesoblastic nephroma (CMN), the most common renal tumour in neonates. We present a case of a full-term neonate born with a palpable abdominal mass, diagnosed as the cellular subtype of CMN. Typically benign, CMN often appears as a symptomless abdominal mass without cystic elements. The neonate underwent a radical left nephroureterectomy, and a histopathological examination confirmed the diagnosis.

Despite its generally benign nature, CMN can lead to serious complications, necessitating early surgical intervention. Our case, 18 months post-operation, shows no evidence of recurrence, underscoring the importance of regular surveillance in managing this rare renal tumour in neonates. This review discusses the diagnostic modalities, subtypes, genetic implications, and varying clinical outcomes associated with CMN, contributing to the understanding and management of this rare pediatric condition.

Keywords: *Congenital mesoblastic nephroma (CMN), nephrectomy, cellular CMN, ETV6, infantile tumour.*

INTRODUCTION

Renal tumours are a significant type of abdominal tumour observed in infants and children, with Wilms tumour (WT), also known as nephroblastoma, being the most common, followed by neuroblastoma [1]. A broad spectrum of renal tumours, ranging from benign to extremely malignant tumours, have been observed. The diagnosis of renal tumours in children typically occurs at a mean age of 36 months, with the majority of cases presenting between 12 and 48 months [2]. Interestingly, approximately 20% of all renal tumours in children can manifest before the age of 6 months. Neonatal renal lesions are infrequent and may include benign and malignant lesions.

Congenital Mesoblastic Nephroma (CMN) has a high prevalence among renal tumours presenting in the perinatal period, comprising over 50% of cases, while WT, rhabdoid tumour of the kidney, and clear cell sarcoma are the subsequent tumour types. However, it is important to note that beyond three months of age, CMN represents less than 10% of all renal tumours [3].

The colour and consistency of CMN inherently resemble a uterine fibroid, exhibiting a homogenous appearance. The histopathological variants include cellular type (66%), classic type (24%), and mixed type (10%). CMN is categorized as a benign tumour and can effectively be treated by nephrectomy alone, although it is important to ensure generous margins are included around the tumour to prevent local recurrence [4].

**Corresponding author: Hina Yousuf, Liaquat National Hospital and Medical College, Karachi, Pakistan, Email: henahyousuf@gmail.com*
Received: April 04, 2024; Revised: June 14, 2024; Accepted: June 29, 2024
DOI: https://doi.org/10.37184/jlnh.2959-1805.2.19

We present a case of a full-term neonate born in our hospital to a 30-year-old female. The newborn was born with a distended abdomen and a palpable abdominal mass. It was subsequently confirmed histologically to be a cellular subtype of congenital mesoblastic nephroma.

CASE REPORT

Following a 38-week gestation period, the male child was born *via* emergency cesarean section. The mother's pregnancy progressed smoothly, with all antenatal scans documenting normal findings. The child was born with a birth weight of 3000 g and a good APGAR score. He passed meconium within 24 hours of birth and tolerated feeding. No voiding issues were noticed.

His examination of the abdomen revealed a disproportionate shape. Palpation showed a bimanual palpable mass in the left upper quadrant, approximately measuring 8 x 6 cm, and had a firm texture with no tenderness or mobility. It had regular margins, and the skin over it was not fixed to the mass and exhibited a normal colour and contour.

The hematologic workup was within normal limits. An abdominal ultrasound revealed a mass on the left renal side, measuring 6.16 x 5.78 x 6.71 cm, exerting pressure on the residual renal tissue in the apical region. Conversely, the right kidney displayed no structural abnormalities. An abdominal CT scan with IV contrast confirmed the presence of a homogeneous soft tissue mass involving the lower and mid poles of the left kidney, enveloping the aorta and left renal vessels (**Fig. 1**). The rest of the scan was unremarkable.

Following the patient's history, examination, and radiological evidence a contingent diagnosis of CMN

  ISSN: 2960-2963 (Online) *Journal of Liaquat National Hospital 2025; 3(1): 64-67*

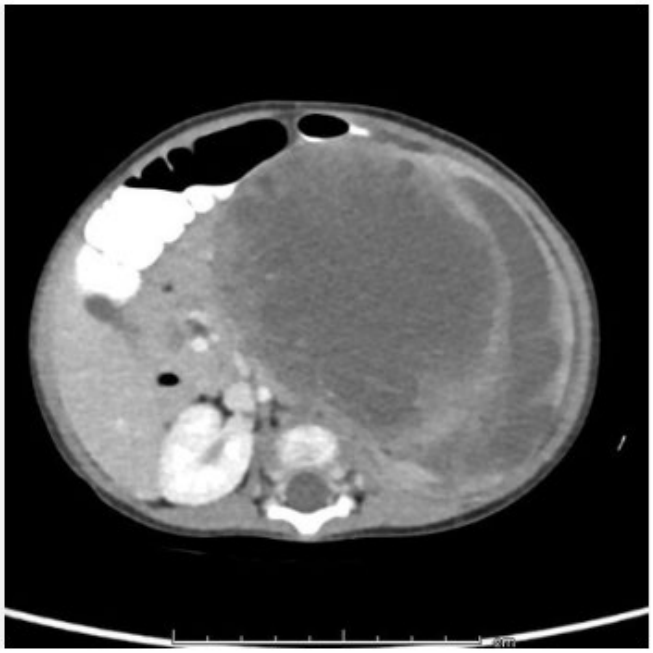

Fig. (1): CT scan with IV contrast revealed a soft tissue mass that involved the left kidney enveloping the aorta and left renal vessels.

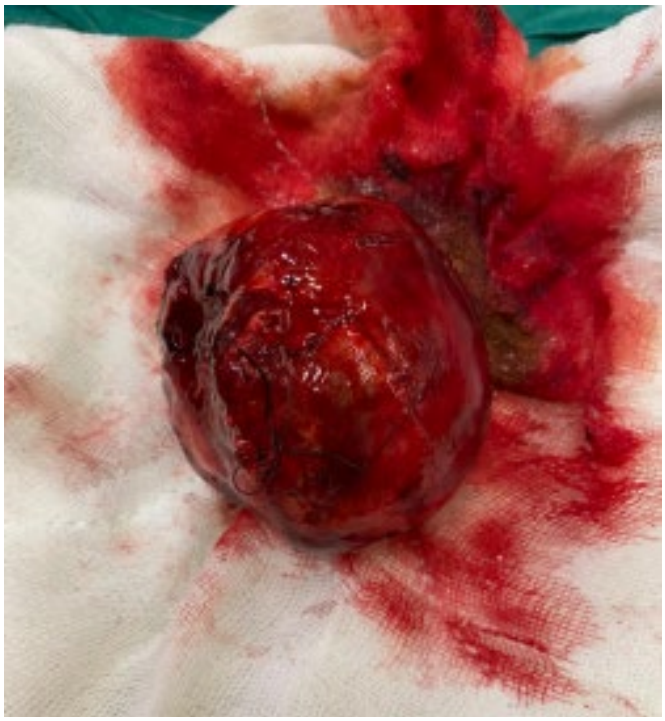

Fig. (2): Showing mass resected completely and distal ureter tied with vicryl.

was proposed keeping WT as a potential differential. Upon exploration, a firm mass originating from the lower pole of the left kidney was found in the retroperitoneum having flimsy adhesions with the peritoneum and perinephric fat. Hilar vessels and the IVC appeared normal. The left kidney appeared normal. He underwent a radical left nephroureterectomy (**Fig. 2**). The mass was resected in completion and the distal

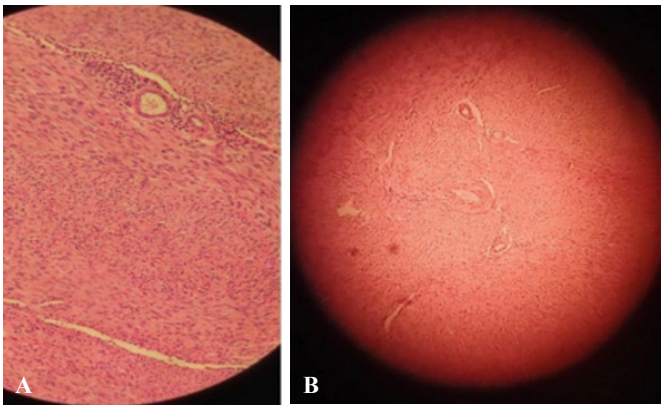

Fig. (3): (A) and **(B):** Slides block showing spindle cells arranged in the form of interlacing fascicles, the spindle cells have elongated nuclei surrounded by pale eosinophilic cytoplasmic borders.

part of the ureter was tied with a vicryl suture before division. The contralateral kidney and the liver were normal on inspection.

Examination of sections obtained from nephrectomy specimens demonstrated distinct features of mesoblastic nephroma. Immunohistochemistry was employed to provide additional evidence supporting the specific diagnosis. The gross size of the excised kidney measured 7x6x6 cm cut sections and composed of spindle cells arranged in the form of interlacing fascicles, the spindle cells have elongated nuclei surrounded by pale eosinophilic cytoplasmic borders. The tumour margin reached up to the outer painted margin infiltrating the pelvis region, renal sinus fat and perirenal fat. The ureteric resection margin was found to be involved in the tumour. Immunohistochemistry coupled with microscopic features suggestive of a cellular variety of CMN (**Fig. 3A&B**).

DISCUSSION

CMN, also known as leiomyomatous hamartoma, is a common condition that often manifests in infants before the age of six months. It accounts for around 3% of all kidney tumours in newborns and is often benign [1]. It is an uncommon tumour that manifests as 1 in 25,000. The differential diagnosis for solid renal tumours includes CMN, rhabdoid tumour, hamartomas, clear cell sarcoma, and nephroblastoma (WT) [2].

First described as a renal tumor of infancy by Bolande *et al.*, [5] in 1967, the tumor is histologically subdivided into three subtypes classical, cellular and mixed. The classical subtype (24%) is the more benign one, whereas cellular (66%) and mixed (10%) variants show a more aggressive pathology with recurrent or metastatic diseases occurring in these subtypes [3]. These have been identified based on mitotic activity and cellularity. In the cellular subtype of CMN, there is production

of the ETV6-NTRK3 fusion-gene product which is a fusion also found in congenital fibrosarcoma, another type of pediatric mesenchymal neoplasm [4].

In infants, CMN frequently develops as an asymptomatic abdominal mass, which on examination seems to be a hard solid mass devoid of cystic components. The tumour has uneven margins. Following this presentation, hypertension, hypercalcemia, and hematuria are the additional usual findings [1]. Diagnosis can be made prenatally by imaging techniques like ultrasounds, CT scans and MRI, however, the best test is a histological study [6]. Generally, CMN has a good prognosis, though, in 14% of cases, serious complications can be associated with this tumour such as polyhydramnios, preterm labour, respiratory distress syndrome, hydrops fetalis, and the development of metastases [7].

CMN has a high curability rate that requires early surgical intervention like complete surgical resection of the kidney and ureter in the first week of life. After tumor resection, infants should be followed clinically and radiologically for 18 months, to detect any recurrence [7]. The first case which was identified prenatally indicating a CMN in a neonate was reported in 1985 by Howey *et al.*, [8] where ultrasound imaging revealed an abdominal tumour in a 28-week-old fetus with polyhydramnios. After the infant's delivery, the lump was surgically removed, and it was later histologically discovered to be a congenital mesoblastic nephroma. This early study lays the foundation for the prenatal identification of the cases of CMN and with the advancement of techniques in the present era, it should be ideal to have identified the conditions manifesting as abdominal mass [8].

Disease relapse occurring as either local recurrence or metastasis to liver, bone and lung within a year of primary resection is observed in 4% of the patients [9]. Mata *et al.* mentioned in their case report of an infant who after undergoing a right nephroureterectomy on the 10th day of life, reported microcephaly, hypotonia, cerebral atrophy and tetraparesis within the first year of life [10]. However, another case by Whittle *et al.*, mentions complete resection by uncomplicated left nephrectomy in an 8-month-old female who showed no recurrence even though the tumour was untreated for 8 months [11]. Current or metastatic diseases occurring in these subtypes have been identified based on mitotic activity and cellularity. In the cellular subtype of CMN, there is production of the ETV6-NTRK3 fusion gene product, which is also found in congenital fibrosarcoma, another type of pediatric mesenchymal neoplasm. Our patient is currently 18 months post-operative and there is no evidence (clinical and radiological) of recurrence. We plan to continue 6 monthly surveillance with ultrasound.

CONCLUSION

This case report highlights the effective management of congenital mesoblastic nephroma (CMN) in a neonate through timely surgical intervention. The favourable prognosis typically associated with CMN was observed, as evidenced by the absence of recurrence 18 months post-operation. This underscores the critical role of vigilant postoperative surveillance in ensuring long-term positive outcomes.

CONSENT FOR PUBLICATION

Written informed consent was taken from the participants.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

ACKNOWLEDGEMENTS

Declared none.

REFERENCES

1. Anakievski D, Ivanova P, Kitanova M. Laparoscopic management of congenital mesoblastic nephroma- case report. Urol Case Rep 2019; 26: 100979.
DOI: https://doi.org/10.1016/j.eucr.2019.100979 PMID: 31388496

2. Pachl M, Arul GS, Jester I, Bowen C, Hobin D, Morland B. Congenital mesoblastic nephroma: a single-centre series. Ann R Coll Surg Engl 2020; 102(1): 67-70.
DOI: https://doi.org/10.1308/rcsann.2019.0111 PMID: 31508997

3. Tripathy PK, Behera S, Mohanty HK. Cellular Congenital Mesoblastic Nephroma in a Newborn. J Neonatal Surg 2017; 6(2): 45.
DOI: https://doi.org/10.21699%2Fjns.v6i2.564 PMID: 28770142

4. Anderson J, Gibson S, Sebire NJ. Expression of ETV6-NTRK in classical, cellular and mixed subtypes of congenital mesoblastic nephroma. Histopathology 2006; 48(6): 748-53.
DOI: https://doi.org/10.1111/j.1365-2559.2006.02400.x PMID: 16681692

5. Bolande RP, Brough AJ, Izant Jr. RJ. Congenital mesoblastic nephroma of infancy. A report of eight cases and the relationship to Wilms' tumor. Pediatrics 1967; 40(2): 272-8.
PMID: 4378178

6. Liu T, Al-Kzayer LFY, Sarsam SN, Chen L, Saeed RM, Ali KH, *et al.* Cellular congenital mesoblastic nephroma detected by prenatal MRI: a case report and literature review. Transl Pediatr 2022; 11(1): 163-73.
DOI: https://doi.org/10.21037/tp-21-289 PMID: 35242663

7. Tongsong T, Palangmonthip W, Chankhunaphas W, Luewan S. Prenatal course and sonographic features of congenital mesoblastic nephroma. Diagnostics (Basel) 2022; 12(8): 1951.
DOI: https://doi.org/10.3390/diagnostics12081951 PMID: 36010301

8. Howey DD, Farrell EE, Sholl J, Goldschmidt R, Sherman J, Hageman JR. Congenital mesoblastic nephroma: prenatal ultrasonic findings and surgical excision in a very-low-birth-weight infant. J Clin Ultrasound. 1985; 13(7): 506-8.
DOI: https://doi.org/10.1002/jcu.1870130716 PMID: 2997304

9. Grosinger L, Salik I, Mehta B. Infantile congenital mesoblastic nephroma leading to multi-systemic end-organ disease. Cureus 2022; 14(10): e30513.
DOI: https://doi.org/10.7759/cureus.30513 PMID: 36415355

10. Mata RP, Alves T, Figueiredo A, Santos A. Prenatal diagnosis of congenital mesoblastic nephroma: a case with poor prognosis. BMJ Case Rep 2019; 12(8): e230297.
DOI: https://doi.org/10.1136/bcr-2019-230297 PMID: 31466985

11. Whittle S, Gosain A, Brown PY, Debelenko L, Raimondi S, Wilimas JA, *et al.* Regression of a congenital mesoblastic nephroma. Pediatr Blood Cancer 2010; 55(2): 364-8.
DOI: https://doi.org/10.1002/pbc.22486 PMID: 20582939