

Congenital Mesoblastic Nephroma: A Case Report

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Abstract

Background: Congenital Mesoblastic Nephroma (CMN) is a mesenchymal renal tumour of early life. With a median diagnosis of two months and over 90% of cases occurring within the first year of life, it is the most prevalent non-Wilms' renal tumour. Even though imaging can be used as a diagnosing tool, it is frequently identified in the neonatal period when the baby has an abdominal mass. There are three different histologic types: mixed, cellular, and classic. Radical nephrectomy is the usual mode of treatment, though adjuvant chemotherapy may be necessary for the cellular type, which may be aggressive. **Case presentation:** We report on a case of a 2-day-old term male neonate born to a 27-year-old mother through spontaneous vertex delivery (SVD). He presented with a right-sided abdominal mass from birth. An abdominal ultrasound scan revealed a huge, well-circumscribed heterogeneous soft tissue mass in the right hemi-abdomen extending to the left side, measuring 10.2 cm by 8.0 cm. He underwent a right radical nephrectomy with a tumour weight of 450 g and a size of 18 cm × 15 cm × 6 cm. Histopathological diagnosis was Congenital Mesoblastic Nephroma (cellular type) Stage 1. **Conclusion:** Any infant with a renal tumour should be evaluated for congenital mesoblastic nephroma. Detailed investigation and complete resection are fundamental for ensuring an excellent outcome.

Keywords

Congenital, Mesoblastic Nephroma, Nephrectomy, Neonate

1. Introduction

Congenital mesoblastic nephroma (CMN), a benign mesenchymal renal tumour,

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accounts for 2% - 3% of renal tumours in children and is the most prevalent renal tumour in neonates and infants under six months of age [1]. Three pathogenic variations of CMN exist: the mixed form, the more aggressive cellular CMN, and the classic CMN [1]. The prognosis for classic CML is generally good. However, cellular CML carries a risk of malignancy and can metastasize and recur [2]. However, if full resection is achieved, surgical resection combined with nephrectomy is seen as an appropriate therapy for all subtypes.

2. Case Report

2.1. Clinical Presentation

This two-day-old term Nigerian male baby, born to a 27-year-old mother, was referred from a Primary health centre on account of the paleness of his body after delivery, vomiting noticed shortly after birth, and abdominal distention noticed on the second day of life. Vomiting was precipitated by feeds, and he had about three episodes for the first two days. Vomitus was initially mixed with altered blood but cleared by the next day. The abdominal distention was noticed by his grandmother, necessitating their presentation at a Primary Health centre. The pregnancy was supervised at a health facility, but the mother also sought the services of a Traditional Birth Attendant (TBA), where she received herbal-based mixtures during the last three months of her pregnancy. Two prenatal scans done at the fourth and eighth months of pregnancy were said to have been normal. Labour was prolonged, and delivery was spontaneous vertex delivery by a TBA at term. He cried after five minutes of resuscitation and weighed 4 kg at birth.

Examination findings were severe pallor and respiratory distress. He had a respiratory rate of 51 cycles per minute, a heart rate of 180 beats per minute, a blood pressure of 73/30 mmHg, a temperature of 37°C and SPO₂ of 97% in room air. His abdomen was distended and tense, with visible anterior abdominal wall veins. An abdominal girth of 36 cm was measured 5 cm from the xiphisternum, and a firm mass was felt in his right lumbar region. Other abdominal organs were difficult to palpate. Other systemic examination findings were essentially normal. There were no clinical signs suggestive of co-existing congenital anomalies. A provisional diagnosis of Wilms tumour was made.

2.2. Laboratory and Radiologic Findings

Blood investigations reported a Packed Cell Volume of 11%, white cell count of $22.4 \times 10^9/L$, platelets of $134 \times 10^9/L$, creatinine was 180 $\mu\text{mol/L}$ and urea of 12.6 mmol/L.

An abdominal ultrasound scan showed a huge, well-circumscribed heterogeneous soft tissue mass in the right hemi-abdomen extending to the left side, measuring 10.2 cm by 8.0 cm. Doppler interrogation showed internal and peripheral flow within the mass. The right kidney was not clearly visualised. The left kidney was normal in size (3.6 cm \times 1.9 cm), position and echotexture with good cortico-medullary differentiation (Figure 1 & Figure 2).



Figure 1. Ultrasound findings showing a normal left kidney vs a right well-defined heterogenous mass.



Figure 2. Ultrasound findings of cystic areas within the mass and displacement of adjacent structures.

2.3. Surgery

A large lobulated right kidney was discovered intraoperatively. A tentative diagnosis of Congenital Mesoblastic Nephroma was made, with Wilms' tumour as the differential. The liver, pelvic organs, and contralateral kidney were all grossly normal. She recovered from the surgery without any complications, and her urine production was normal. The patient had a total right nephrectomy on the twelfth day of life.

2.4. Pathology

2.4.1. Gross Anatomy

The mass was sent for histologic analysis and was grossly described as soft to firm tissue with a multilobulated external surface. A 15 × 13 cm fleshy tumour with a greyish-white colour was visible on the sliced surface. There were many cysts within the partially solid remnant of renal tissue, the largest of which measured 1 cm by 0.7 cm and contained serous fluid. Areas of the solid surface were necrotic. No lymph nodes or perinephric fat were seen (**Figure 3** & **Figure 4**).

2.4.2. Microscopy

Histology sections of the kidney show an unencapsulated cellular tumour consisting of spindle-shaped cells as intermingling fascicles with few foci of collagen deposition. These cells have a moderate amount of cytoplasm with plump vesicular to hyperchromatic nuclei that are mildly pleomorphic, with brisk mitosis seen. A

clear boundary exists between the tumour cells and the normal kidney tissue. The renal sinus is free of tumour cells.

A histological diagnosis of *CONGENITAL MESOBLASTIC NEPHROMA (CELLULAR TYPE) STAGE 1* was made (**Figure 5** & **Figure 6**).



Figure 3. Gross picture of the right renal mass with an external greyish white to tan multi-lobulated surface.

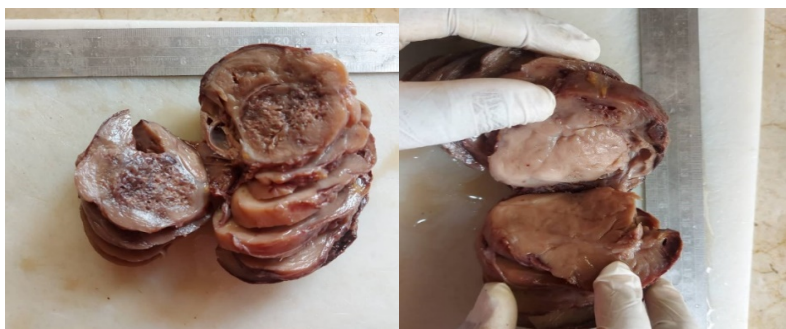


Figure 4. The cut surface of the kidney showing a greyish-white fleshy tumour and necrotic areas.

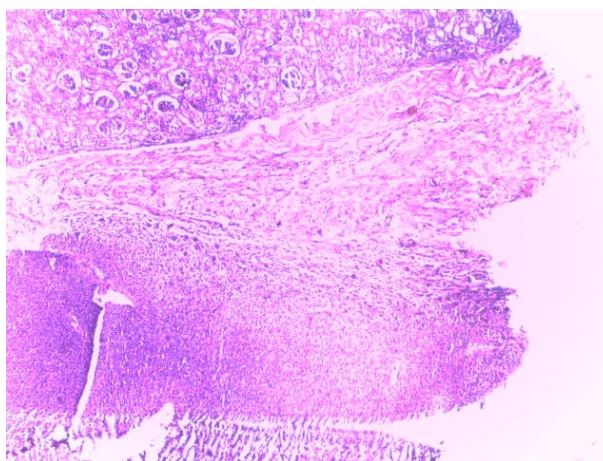


Figure 5. Section showing demarcation of tumour from normal-appearing renal parenchyma ($\times 40$).

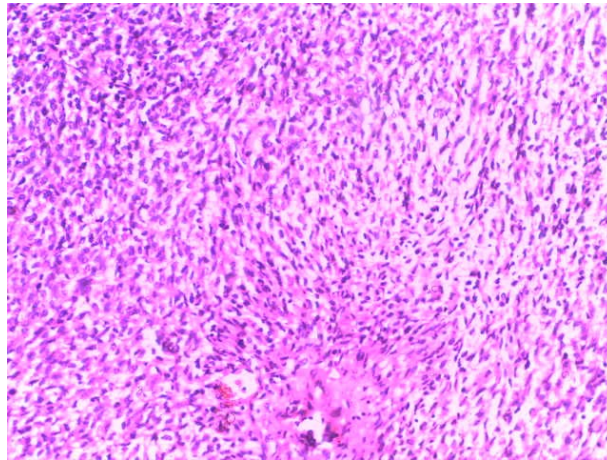


Figure 6. Section showing fascicles of spindle-shaped proliferation with collagen deposition ($\times 200$).

2.5. Follow-Up

He has been and is still on multidisciplinary follow-up with the Paediatric Oncologists, Paediatric Nephrologists and Paediatric surgeons. He has no complaints; he has normal growth and no problems with development. A repeat abdominal ultrasound scan three months after surgery reported normal findings with no features of tumour recurrence. His last hospital visit at seven months of age showed him to be stable with no evidence of metastasis.

No other accompanying anomalies have been identified. Karyotyping and genetic studies were requested for possible aneuploidies which is seen in a lot of cases of CMN, however these are yet to be done due to logistic reasons.

3. Discussion

Congenital Mesoblastic Nephroma (CMN) is a mesenchymal renal tumour of early life. With a median diagnosis of two months and over 90% of cases occurring within the first year of life, it is the most prevalent non-Wilms' renal tumour [2]. Up to 90% of cases are found by the time the child is a year old, with the majority of diagnoses occurring in the first three to six months of life [3] [4]. The right kidney-to-left kidney ratio is about 1:1, while the male-to-female ratio is approximately 1.5:1 [5]. Three subtypes have been identified: mixed, cellular, and classic. As seen in our patient demonstrated, the cellular type is more prevalent than the other two. The cellular type (66%) resembles infantile fibrosarcoma, the classical type (24%) resembles infantile myofibromatosis, and the mixed type (10%), which is a mixture of the two, resembles composite fibromatosis [1]. A greater number of cases are anticipated to be detected before delivery as prenatal ultrasonography becomes more widely used [6].

Congenital mesoblastic nephroma, Wilms' tumour, and considerably less common lesions such as malignant rhabdoid tumour, clear cell sarcoma of the kidney (CCSK), and ossifying renal tumour of infancy are among the differential diagnoses for solid renal tumours [2]. While CMN is more common in younger infants,

Wilms' tumour (WT) is the most common paediatric renal tumour and might be mistaken for cellular CMN at imaging. Another congenital tumour that may invade the kidney, typically the upper pole, is neuroblastoma. It very seldom develops within the renal parenchyma. It can be identified by its propensity to enter the spinal canal, encase vessels, and cross the midline [7].

Making a differential diagnosis between WT and CMN is essential to creating the best possible treatment plan. According to an analysis of imaging features and clinical symptoms, WT is similar to CMN, especially the cellular form; however, less than 2% of patients with WT present before the age of three months. Cancers with bilateral growth and congenital syndromes or abnormalities are more suggestive of WT. In-depth morphological data from electron microscopy can be used to make the necessary differential diagnosis.

Although CMN is often a benign tumour, there is a chance of local recurrence and distant metastases, with the lung being the primary location of metastasis. Other sites, including the liver, bone or brain, have been reported in 5% - 10% of cases. Total radical nephrectomy is curative in most patients [3]. Wide surgical margins are necessary because of the infiltrating boundaries and propensity to infiltrate the perinephric fat and hilum. Incomplete resection may result in local recurrence. Chemotherapy and other multimodal therapies are therefore reserved for cancers that cannot be surgically removed or that have residual tumours [4].

It is advised to have routine follow-up abdominal ultrasonography scans. In our case, the tumour was completely resected following a radical nephrectomy. A repeat abdominal ultrasonography scan at three months of age revealed normal findings, with no evidence of metastasis; therefore, an excellent recovery is anticipated. He is currently being followed up by the oncology unit of the hospital. Follow-up visits have been regular at monthly intervals and he is in stable condition as at seven months of age.

4. Conclusion

In infants, CMN is the most prevalent type of renal tumour and is often benign. Since it can be identified early in pregnancy, it is advisable to promote foetal anomaly screening using ultrasound to prevent late presentation. While CMN is usually benign, patients must undergo routine monitoring for any possible paraneoplastic syndromes. Proper follow-up is necessary in the first few years of life, because certain forms of mesoblastic nephromas can potentially develop into malignancies.

Consent

Informed written consent was obtained from the patient's parents. This report, however, does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current criteria for Authorship.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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