IBOM MEDICAL JOURNAL
Vol.18 No.3 | July - September 2025 | Pages 556-558
www.ibommedicaljournal.org

Congenital mesoblastic nephroma: A case report and review of literature

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Abstract

Introduction: Congenital mesoblastic nephroma is a rare renal tumour affecting newborns and infants. It is also known as leiomyomatous or fetal hamartoma and is the most common solid Renal tumor in the neonate and infants less than 6 months old.
Case summary: We report a case of congenital mesoblastic nephroma (CMN) in a 19-hour-old boy presenting with a huge intra-abdominal mass and failure to pass meconium. Ultrasound scan revealed a huge intra-abdominal mass arising from the left renal bed and gaseous intestinal dilatation. Emergency simple total left nephrectomy was done, and histological diagnosis of CMN was confirmed. The newborn was discharged in good condition 5 days after surgery.
Conclusion: A high index of suspicion and prompt surgical intervention are crucial for a favourable outcome.

Key words: congenital, mesoblastic nephroma, emergency surgery

Introduction

Congenital mesoblastic nephroma is a rare paediatric renal tumour affecting mainly neonates and infants, with 90% of the tumours being diagnosed within the first year of life, and is associated with polyhydramnios and premature delivery[1], It rarely occurs after 3 years of age even though few have been reported in adult[2]. It comprises mainly two histological subtypes, classic and cellular, with the latter accounting for two-thirds of cases[3]. Mixed variant has also been reported. It poses a diagnostic challenge to both surgeons and pathologists due to its rarity and similarity with other, more common tumours such as Wilms' tumour. This tumour generally has a favourable prognosis[4]. We aim to report this rare neonatal tumour and to heighten awareness of its ability to cause neonatal intestinal obstruction, which is a rare occurrence.

Case Report

A 19-hour-old male neonate was referred to our facility with a provisional diagnosis of intestinal obstruction presenting with failure to pass meconium, difficulty in breathing, and a grossly distended abdomen. He was a term neonate delivered to an 18-year-old primipara. Pregnancy was booked at 5 months ' gestational age for antenatal care (ANC) in a primary health care centre. She has been regular with her ANC visits, but no ultrasound scan (USS) was done throughout the pregnancy. The delivery was at term, and it was uneventful, though the baby did not cry immediately after delivery. On examination, he was in mild respiratory distress with a respiratory rate of 65 cycles per minute, heart rate of 160 beats per minute, and normal heart sounds, with no murmurs. The SPO_2 was 95% on atmospheric air. He was globally pink,

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DOI: 10.61386/imj.v18i3.718

anicteric, with a grossly distended abdomen that made it difficult to palpate abdominal organs. The anal opening was normally sited with empty rectum. Abdominal USS showed a huge, well-circumscribed mixed echogenic mass measuring about 6.4 X 7.6cm occupying the whole abdominal cavity but more to the left side of the abdomen. Plain abdominal X-ray shows gaseous distention of stomach and bowel loops. No area of calcifications was noted. The packed cell volume (PCV, full blood count (FBC), and serum urea & electrolytes are all within normal ranges. An emergency exploratory laparotomy was then performed, and a huge left renal mass about 10cm X 8cm in its widest dimension was noted pushing and compressing the loop of dilated bowel. Aorta, inferior vena cava, and the renal vessels were not displaced, and they were free. Left simple total nephrectomy was performed, see Figure 1. The specimen was sent for histological diagnosis.

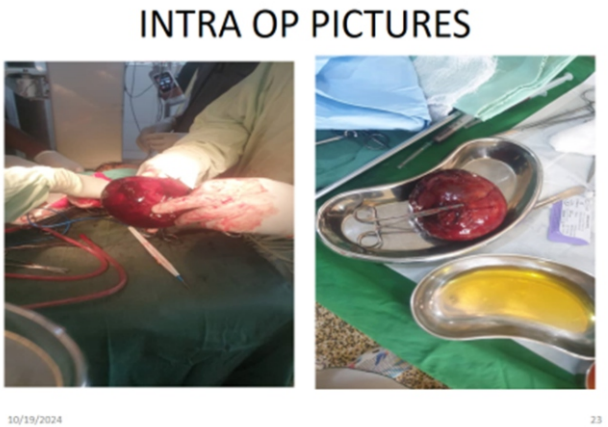

Figure 1: intraoperative picture showing the tumour (A); Resected specimen showing the tumour in a kidney dish (B)

The patient did not have any postoperative complications and was discharged on the 5th day post op. He was subsequently seen for follow-up with no complaint.

Grossly, the mass was relatively well circumscribed with a greyish white surface with areas of haemorrhages. An area of dilated renal sinus was noted. Histologically, cellular mesenchymal proliferation composed of plump spindle cells disposed in diffuse sheet-like growth with minimal atypia and rare mitotic figures was noted. Few foci of extramedullary haematopoeisis were noted with focal invasion of the renal sinus. The overall histological features were those of Congenital Mesoblastic Nephroma, cellular type, stage II (due to renal sinus invasion). See Figure 2.

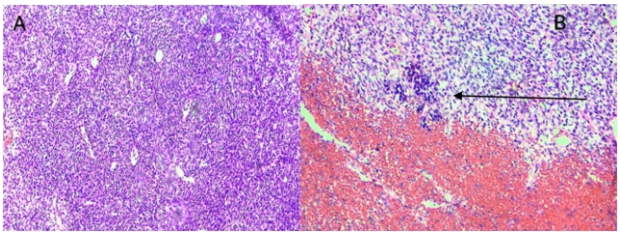

Figure 2: Histological sections showing a cellular but blad proliferation of ovoid to spindle cells interspersed by thin-walled vessels (A); areas of haemorrhages and extramedullary haematopoeisis (arrow) is also noted (B) H & E X 200

Discussion

Congenital abdominal masses as well as other overt congenital abnormalities are commonly diagnosed prenatally in recent times due to advances in antenatal care, especially in the developed world[1,5]. In low- and middle-income countries, such services are still lacking in many communities[6]. In our index patient prenatal ultrasound scan was not done throughout the pregnancy period due to a lack of an ultrasound scanning machine in the primary health care center. Despite widespread use of Ultrasound Scan, only 15% of renal masses are diagnosed prenatally[6]. Congenital mesoblastic nephroma is a rare renal tumour accounting for 3 – 5% of neonatal renal tumours[1,4,7]. It is the most common neonatal renal tumour and is more prevalent than Wilms' tumor in infants younger than 3 months of age[3,8]. Polyhydramnios is observed in almost 70% of CMN cases[9].

The mechanism of polyhydramnios is not clear, but it may be due to intestinal obstruction by the renal mass and polyuria resulting from increased renal perfusion[4,10]. Our patient did not present with a history of polyhydramnios, nor prematurity but presented with features of intestinal obstruction. Our case also represents a rare and complex scenario with a diagnostic conundrum: a 19-hour-old neonate with a huge CMN associated with failure to pass meconium requiring emergency left nephrectomy is noteworthy. CMN can cause extrinsic compression of adjacent organs, leading to intestinal obstruction[11]. This association underscores the importance of considering CMN in neonates with intestinal obstruction and abdominal masses. The decision to perform emergency exploration was crucial due to the intestinal obstruction. Computerised tomographic

scan would have been an investigation modality that would have suggested the diagnosis, but due to the age of the child is not suitable due to high radiation exposure and the emergency nature of the intestinal obstruction. Magnetic Resonance Imaging (MRI) is an essential imaging tool in the diagnosis, treatment planning and follow-up of this tumour. Its high spatial resolution and ability to characterize tissue composition make it an invaluable tool for managing this disease. Our case contributes to the limited literature on this condition in this age group. The case is consistent with reported cases of most CMN, however, the association with intestinal obstruction and emergency nephrectomy is distinct. Classical CMN, confirmed by histology, has a favourable prognosis with a low risk of recurrence and metastasis compared to cellular or malignant variants. Recommended treatment for a patient with cellular CMN includes adjuvant chemotherapy (actinomycin D plus cyclophosphamide). Complete surgical excision appeared curative for infants less than 3 months old with even cellular congenital mesoblastic nephroma and those of all ages with classical congenital mesoblastic nephroma[12]. Our patient had classical CMN with clear histologic margin, and is a day old, so he does not require chemotherapy.

Conclusion

Congenital mesoblastic nephroma is a rare neonatal renal tumour with good prognosis. Treatment is essentially surgical, as in our patient, who was a 19-hour-old male neonate, though Cellular variety may rarely require chemotherapy. Presentation as an emergency case in our patient is distinct, and such should be taken into account if a neonate presents with intestinal obstruction.

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