

Congenital Mesoblastic Nephroma: Case Report

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Abstract:

Congenital mesoblastic nephroma is a rare neonatal renal tumor, comprising of different histologic types. It constitutes 2% of neonatal renal tumors and 10% of renal mass in infants younger than 6 months. Recurrence is higher when they are younger without chemotherapy.

A term neonate with abdominal mass with associated septicemia, had nephrectomy of the right kidney, but represented at 6 months with recurrent tumor.

Pathologist should be encouraged to pay close attention to renal tissues to minimize misdiagnosis and clinician to be mindful of recurrence of diagnosis is made.

Keyword: Neonate , Congenital Mesoblastic Nephroma, Recurrent

Introduction:

Congenital mesoblastic nephroma (CMN) described in 1967 and also called leiomyomatous hamartoma is the most common neonatal renal neoplasm with a distinct entity which should be differentiated from Wilms tumor.¹ It comprises 3%-10% of paediatric tumor. CMN is the most common tumor in children less than 6 months.² Renal masses in neonates are usually benign and CMN constitute 2% of renal masses in the neonates.³ Wigger and Bogdan are of the opinion that the tumor is hamartomatous while Bolande believed the origin is from renal blastema.¹ CMN and infantile fibrosarcoma are likely to represent a single neoplastic entity because of the association with polysomies for chromosome 8,11,17,20⁴ Synder et al in explaining the histogenesis proposed a theory using "two-hit" model that CMN would occur after a neoplastic mutation in early embryogenesis whereas atypical cellular will develop in later stages before blastema

undergoes metanephric differentiation. In both second mutation is therefore necessary to produce malignant transformation. These are aptly supported by immunohistochemical study.⁵⁻⁷ It is not yet possible to identify variability in biological behavior of tumor and this is further strengthened by DNA diploid study where the typical CMN demonstrated diploid and others were aneuploid.⁸ CMN was thought to be initially benign but recent advances have suggested it as a spectrum, incidence of cellular is 42%-63%.⁹ No identifiable predisposing factor.⁸ Incidence is 8 per million in children less than 15 years.¹⁰

Male preponderance has been reported.¹¹ Age at presentation is dependent on the histologic type.² 14% will have associated congenital anomaly.¹² The overall prognosis is good however the course can become unpredictable when present in the perinatal period as well as atypical case.¹³

This patient posed a diagnostic challenge because he was referred as intestinal obstruction probably because of vomiting and abdominal distention, he was however managed for septicemia and Wilms'. CMN was not thought of until recurrence. This case is being reported so that clinically when neonates presents with renal mass this entity should come to mind and chemotherapy should be considered in the protocol of management to prevent recurrence. To the best of our knowledge this is the first case from our centre.

Case report

OA a male neonate product of term gestation was delivered by spontaneous vertex delivery was referred on the 12th day of life with a diagnosis of intestinal obstruction. History is

that of abdominal swelling which was noticed on the 10th day of life, progressive and diffuse with associated fever and vomiting and no hematuria. There was no history of polyhydramnious but mother had a febrile illness associated with vaginal bleeding in the first trimester. At presentation he had a temperature of 40c, weight of 3.1kg. right -sided abdominal mass measuring 8cm by 10cm was found on examination. Blood pressure was 80/50mmHg.

He was subsequently managed for neonatal septicemia and renal mass. There was neutrophilic leucocytosis, electrolyte urea and creatinine were normal.

He had right nephrectomy on the 24th day of life (pix1) measuring 11cmx10cmx6cm and histology of the mass was reported grossly as renal mass with areas of nodularity and tumor involving over 90% of kidney, some areas are cystic hemorrhagic infarctions and microscopy revealed poorly circumscribed neoplasm consisting of sheets and nests of proliferating spindle cells, cigar shaped nuclei, infiltrating renal parenchymal. Focal areas showing abortive glomeruli. Loose stroma containing adipose tissue and cystic cavities lined by flattened cells. It therefore concluded it was mesoblastic nephroma. This was repeated at another pathology laboratory after an initial diagnosis of nephroblastoma was made.

Patient there-after remained well till 6th month of life when he presented again with recurrence of abdominal swelling, a repeat scan (cost of which was paid by the author) confirmed the presence of cystic swelling and parents refused fine needle biopsy and was subsequently lost to follow up as parents declined further treatment.

Discussion

The diagnosis of CMN may be made antenatally when there is polyhydramnious as reported in 71% of cases¹⁴ On renal imaging a "ring sign" consisting of concentric hyper and hypo-echoic ring pattern on ultra-sound, polyhydramnious

and fetal hydrops both on Renal ultrasound (RUSS) and magnetic resonance imaging (MRI) is seen in the typical intra renal CMN prenatally.¹⁵

The diagnosis might have been missed in utero either because there was no ultra sound done during pregnancy or the mass that is operator dependent was missed as opined by Lisa et al.¹⁶ Pre operative diagnosis is difficult. Non availability and affordability by the parents limited other investigation like MRI and contrast enhanced computerised tomography as well as identification of immune-reactive markers made diagnosis more difficult.

MRI is the most accurate diagnostic tool depicting the local and regional extension of the mass though confirmation is still histology. These facilities were not available at our center.

Congenital mesoblastic nephroma presentation is dependent on the type, the classic type will usually present within 16 days of life, cellular at 5 months and the mixed at 2 months. The age at presentation of our patient would suggest the classic type and the reason for recurrence as reported by Futwaegner et al (2). Pix 2 shows the leiomyomatous appearance of the tumor.

It has also been found recently by Brian et al that there is similarity in histologic appearance of CMN and infantile fibrosarcoma and are likely to represent a single neoplastic entity because of the association with polysomies for chromosome. Chromosome analysis however was not done in this patient⁴

The differential diagnosis of this tumor will include multicystic nephroma, cystic partially differentiated nephroblastoma and multicystic dysplastic kidney¹⁷ Multicystic nephroma, a type of cystic nephroma has diagnostic criteria which includes unilateral involvement, solitary lesion, multilocular nature, non communication of cysts with one another, loculi lined by epithelium and normal renal tissue when present^{17,18}

The gross appearance of this tumor will make one consider multicystic nephroma but histology differentiates them thus emphasizing the need for histology in the diagnosis though this too has its challenges. The presenting blood pressure was normal, he would have benefitted from long term follow up for detection of hypertension and proteinuria because of the nephrectomy but was unfortunately lost to follow up.

Hyper-calcemia has been suggested as a possible mechanism for polyhydramnios seen antenatally and as a paraneoplastic syndrome linked to hypertension but this could not be confirmed in this patient because serum calcium was not documented. Many patients have been reported to do well after nephrectomy.

Recurrence within first year post surgery without chemotherapy as occurred in this child has been reported by Lina et al¹⁹ and this was possibly why this child represented as chemotherapy was not included in the management, though attempt had been made by the use of flow cytometry to determine the extent of management this was not conclusive⁸ Recurrence is higher within the first year of life, rupture during resection, positive tumor margin, histologic cellular subtype and age. Recurrence in this patient will suggest the cellular type that is aggressive.⁴

Challenges faced in the management of this patient included a misleading diagnosis at point of referral as intestinal obstruction, poor investigation due to lack of funds by the parents and non availability of facility for such. Therapeutically the patient was not offered chemotherapy at first presentation and this may probably have discouraged the parents when tumor recurred.

Conclusion:

Neonatologist should be aware of this entity in neonates and chemotherapy should be given as

an adjunct to prevent recurrence. Pathologists should be encouraged to pay more attention to kidney tissues to minimize misdiagnosis.

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