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Prevalence of Congenital Mesoblastic Nephroma in North West Rajasthan: A five year retrospective study at our institute.

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Abstract

Background: Congenital Mesoblastic Nephroma, while rare, is the most common kidney neoplasm diagnosed in the first three months of life or prenatally and accounts for 3-10% of all childhood renal neoplasms. This neoplasm is generally non-aggressive and amenable to surgical removal.

Methods: The medical records of all the children who underwent nephrectomy and specimen sent to the Pathology Department of Sardar Patel Medical College, Bikaner, in last five years were retrospectively analysed.

Results: Out of total 75 patients, 15 patients had renal tumors and 1 (7.1%) patient had histological evidence of Congenital Mesoblastic Nephroma. The patient was 10 months old female. Nephrectomy was performed.

Conclusion: Congenital Mesoblastic Nephroma is a rare congenital renal tumour and mostly diagnosed within three months of birth.

Keywords: Congenital Mesoblastic Nephroma, Wilm'stumor

Introduction

Renal tumors are infrequently diagnosed in infants younger than 6 months of age [1,2], and neonatal renal tumors account for only 7% of all neonatal tumors[1].Wilmstumor(WT) or Nephroblastoma constitutes the prototypical example of a neoplastic process. It is seen primarily in infants, 50% of the cases occurring before the age of 3 years and 90% before the age of 6 years[3,4]. However, Wilms tumor is only exceptionally seen as a congenital neoplasm, a point of great importance in the differential diagnosis with mesoblastic nephroma.[5]

Congenital Mesoblastic Nephroma (CMN) is a congenital myofibroblastic tumor that resembles infantile fibromatosis/ leiomyoma or fibrosarcoma. It contains fibroblastic cells [connective tissue cells], and may spread to other kidney or to nearby tissue. It is also called fetal mesenchymal/leiomyomatous hamartoma.

Congenital Mesoblastic Nephroma was first named as such in 1967 but was recognised decades before this as fetal renal hamartoma or leiomyomatous renal hamartoma. Bolande who coined the term "Congenital mesoblastic nephroma" described it as nonencapsulated, locally invasive and clinically benign tumor.[6]

It presents as an abdominal mass which is usually detected prenatally by ultrasound and is associated with polyhydroamnios(i.e. excess of amniotic fluid in the amniotic sac), prematurity, hematuria, hypercalcemia and elevated serum levels of renin. It rarely occurs in children older than 2 year of age, however rarely cases have been reported in adults.[5]Most tumor rare located near hilum.[5] It is usually a unilateral tumor and shows slight male predominance.

It is of 3 types:Classical variant (24%) represents benign disease and cellular variant (66%) represents aggressive pathology. There is a mixed variant (10%) having features of both type[7]

Grossly classical variant appears as solid, firm and non-encapsulated mass with irregular margin. Cellular variant has necrosis and large cystic areas and hemorrhage. Mixed variant has cysts, hemorrhage and necrosis in cellular areas.[8]

Microscopically[8], classical variant resembles infantile fibromatosis or leiomyoma with fascicles and whorls of bland spindled myofibroblasts and thin collagen fibers. Tumor surrounds tubules and glomeruli, has irregular borders. Chondroid metaplasia / dysplasia of the entrapped tubules is common. Mitoses are rare and necrosis / desmoplasia are not present.Cellular variant resembles infantile fibrosarcoma with a sheet-like proliferation of plump, atypical spindle cells with abundant cytoplasm, vesicular nuclei and nucleoli; frequent mitotic figures (25 - 30 / 10 HPF) and necrosis. The tumor has a pushing border.Mixed type has tumors with a combination of both classic and cellular.

Immunohistochemistry aids in performing differential diagnosis;CMN generally exhibits the following results: VIM (+), Ki-67 (+), CD34 (−), EMA (−), CK (−), DES (−) and SMA (-)[9]

Congenital mesoblastic nephroma should be differentially diagnosed from Wilm'stumor, congenital infantile sarcoma, Rhabdoid tumor, Clear cell sarcoma of the kidney, Infantile myofibromatosis.

Congenital mesoblastic nephromas lack the abnormalities in chromosome 11 that characterize Wilms tumor and are associated instead with polysomies for chromosomes 8, 11, 17, and 20.[5]

A differential diagnosis between CMN and WT is critical to develop the most effective therapeutic approach. Surgical removal of the entire involved kidney plus the peri-renal fat appeared curative for the majority of all types of mesoblastic nephroma.[10]Removal of the entire afflicted kidney plus the peri-renal fat appears critical to avoiding local recurrences. Therefore, radiation therapy or chemotherapy is not indicated. In up to 7% of the cases, recurrence with local invasion of retroperitoneum will occur and may prove fatal.[11]Cases associated with distant metastases to lung and brain have rarely been reported.[12]

The aim of the present study is to determine the prevalence of Congenital Mesoblastic Nephroma at

Sardar Patel Medical College and associated groups of hospitals, and to compare the prevalence in our region with global and national literature.

Methods

Study design: Retrospective Study.

Study duration: Five years from Janurary 2015 to December 2019.

Study place: Department of Pathology, Sardar Patel Medical College, Bikaner.

Study Material: All specimens of renal biopsy and nephrectomy in children received at Department of Pathology during the study period were taken for study. The clinical and relevant data were recorded from requisition form and patient's clinical records.

The specimens received, were fixed in 10% buffered formalin. Gross examination was done and findings were recorded. After fixation of the tissue, sections were taken as per protocol and processed by wax block method and stained by haematoxylin and eosin stain and observed under light microscope.

Result

We received a total of 75 specimens of renal biopsy and nephrectomy in children at Department of Pathology during the study period. Out of which 15 were of pediatric renal tumors and 1 case (7.1%) was Congenital mesoblastic nephroma. The patient of Congenital mesoblastic nephroma was a 10 days old female child. The tumor was diagnosed prenatally by ultrasound during the third trimester scan, which showed well defined heterogenous hyperechoic mass lesion in right renal fossa of fetus with differential diagnosis of Wilm's Tumor and Congenital mesoblastic nephroma, which was further studied by CECT abdomen postnatally. The patient had undergone right nephrectomy.On gross examination, [Image 1] tumor was solitary occupying almost whole of the renal mass with only a small area of normal renal parenchyma identified and tumor margins with normal kidney were ill defined. Cut surface was grey white, whorled appearance and bulging.Few haemorrhagic and necrotic areas were also seen. On histological examination there were spindle shaped tumor cells resembling fibroblasts presents in sheets and infiltrating between normal renal parenchyma, the features are consistent with Congenital Mesoblastic Nephroma, shown in image 2 [A-D]. The present study is summarised in Table 1.

Table 1: Summary of Present Study

Study duration	Total cases	Neoplastic lesions	Wilm's tumor	Congenital Mesoblastic Nephroma	Prevalence of CMN [of paediatric renal tumors]
Jan. 2015 to Dec. 2019	75	15	10	1	7.1%

Discussion

Congenital mesoblastic nephroma, while rare, is the most common type of renal tumor in newborns and infants under three months of age, and 90% of cases occur in patients under one year old[7].

Congenital mesoblastic nephroma most commonly presents as an abdominal mass, but could also manifest with hypertension or hypercalcemia. The hypercalcemia observed in some infants has been attributed to excessive production of prostaglandin E by the tumor cells[13,14]. Due to rarity of this tumor we are of the view that even this small study will be a useful contribution in determining prevalence of CMN in N-W Rajasthan.

With the advancement of imaging techniques, most CMN are diagnosed prenatally by ultrasonography. However a differential diagnosis between CMN and WT is critical to develop the most effective therapeutic

approach. The clinical symptoms& signs and imaging characteristics shows that WT is similar to CMN in most cases, particularly the cellular variant, but fewer than 2% patients with WT present at under three months of age[7] . Hence histopathological examination of all paediatric renal tumors plays an important role in definitive diagnosis of CMN and to differentiate it with WT to plan treatment properly. The observation in our present study is consistent with the available literature; the patient with CMN was diagnosed with renal tumor prenatally as having right renal mass with differential diagnosis of WT and CMN and treated within three months of life. Likewise none of the cases of Wilm's tumor were diagnosed and treated within 3 months of age.

Congenital mesoblastic nephroma is the most frequent type of renal tumor in the neonatal and early infantile period, comprising 3–10% of all childhood renal tumors. [7] The prevalence of CMN in the present study is reported as 7.1% which align with the available global literature.

Congenital mesoblastic nephroma usually presents as a solid tumor with a smooth, firm or rubbery external surface and a lightly colored or yellow-tan whorled cut surface. Histologically classical variant is ~24%, cellular variant ~66% and mixed variant ~10% [7]. In the present study we reported a single case of CMN with gross and microscopic features suggestive of classical variant (100% of total CMN), which may be due to fewer number of cases in the study.

The prognosis of Congenital mesoblastic nephroma is generally good. Total nephrectomy with removal of perinephric fat is curative in most caese. The 5-year survival and overall survival rates of infants are 94% and 96%, respectively. These rates also depend upon the histological findings. [15]

We compared the finding of present study with other studies conducted in the past. The comparative analysis is shown in Table 2

Table 2: Comparative analysis

S. No.	Study	Study period	Total cases	No. of CMN	Prevalence
1	J C Barrantes et al[16].	1957-1986	211	13	6%
2	Present study	Jan 2015-Dec2019	75	1	7.1%

Conclusion

Congenital mesoblastic nephroma, while rare, is the most common type of renal tumor in newborns and infants under three months of age, which is usually diagnosed prenatally. CMN is often confused with Wilm's tumor, due to similar clinical and imaging findings, so histopathological examination plays an important role in its diagnosis. Nephrectomy with peri renal fat removal is curative in majority of cases.

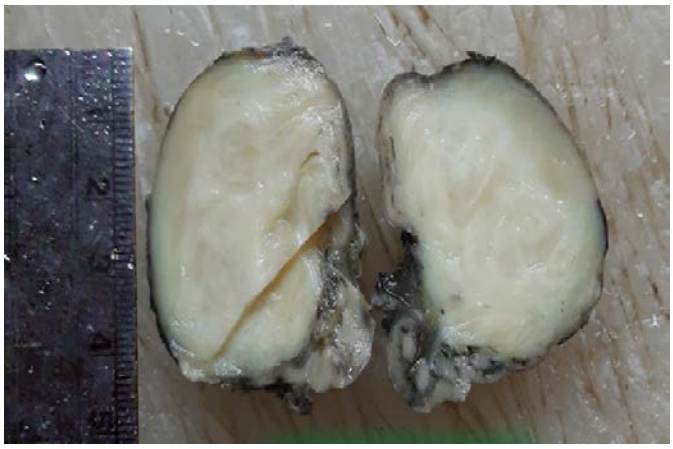

Figure 1: Solitary tumor and cross section is grey white to tan, whorled appearance and bulging. Tumor margins with normal kidney are ill defined.

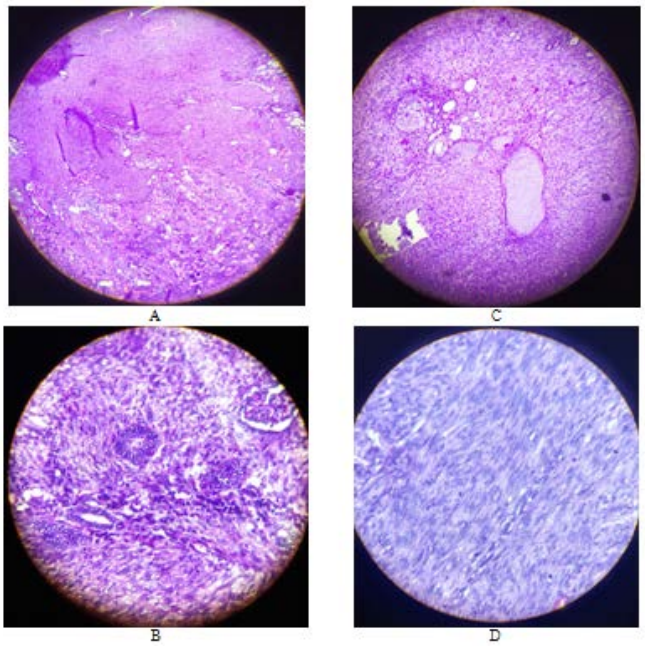

Figure 2:Microscopic examination of H&E stained sections;

[A] 10X Tumor composed of interweaving fascicles of bland spindle cells.

[B] 40X Tumor infiltrates between normal renal structures.

[C]10X Chondroid metaplasia seen within tumor.

[D]40X Spindle cells resemble fibroblasts with mild to moderate nuclear pleomorphism.

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