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Case Report

Congenital Mesoblastic Nephroma (Mixed Type) – A Rare Entity

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Abstract: Congenital mesoblastic nephroma is a mesenchymal renal neoplasm that constitute only about 2-4% of all pediatric renal tumours. But it is the most common neoplasm of neonatal period. Here we present a case of a premature infant girl who was found to have an abdominal mass on physical examination on the first day of life. She underwent Right radical nephrectomy and histopathology revealed Congenital Mesoblastic Nephroma.

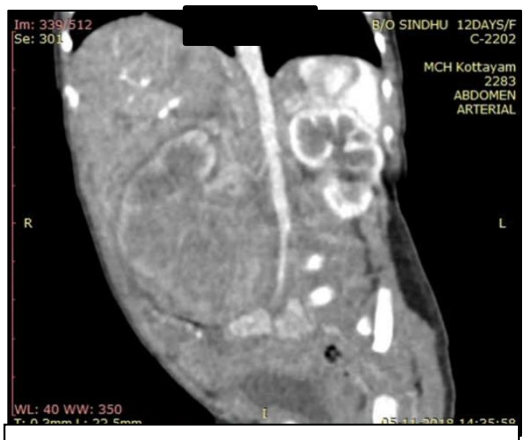
Keywords: Congenital mesoblastic nephroma, mesenchymal renal tumour, neonate.

INTRODUCTION

Congenital mesoblastic nephroma is a mesenchymal renal neoplasm that was described by Bolande in 1967 (Bolande, R.P. *et al.*, 1967). Although CMN constitutes only 2-4 % of all pediatric renal tumours, it is the most common congenital renal neoplasm and 90% of it occurs in the first year of life (Thompson, P., & Chintagumpala, M. (2012). Here we present a case of CMN in a premature baby who presented with a palpable abdominal mass.

CASE REPORT

A premature infant girl was born to a 28 year old woman after 35 weeks of gestation. This baby was the first of the dichorionic diamniotic twins and was delivered vaginally. She had a birth weight of 1.76 Kg and she cried spontaneously after birth. Later she was admitted to NICU for LBW care. Physical examination on first day of life showed a fullness of abdomen and a palpable right flank mass. Her BP was normal. All other blood and urine parameters were normal including Renal Function Tests. Abdominal ultrasonography showed heterogeneous echotexture involving mid and lower pole of kidney. CECT was done after 12 days and it confirmed a heterogeneously enhancing soft tissue density lesion involving lower pole and interpolar region of right kidney. A differential diagnosis of Nephroblastoma or CMN was made.



CECT abdomen

GROSS: cut section whitish whorled

Quick Response Code

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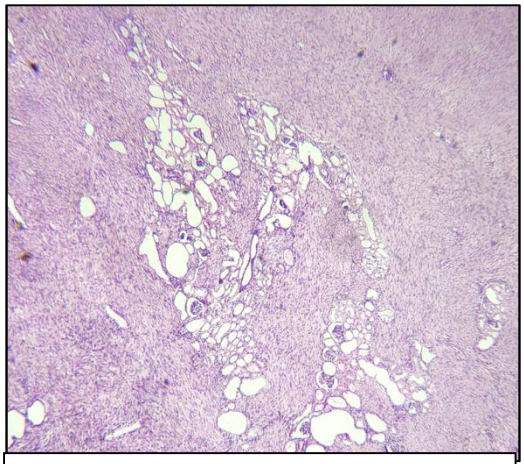
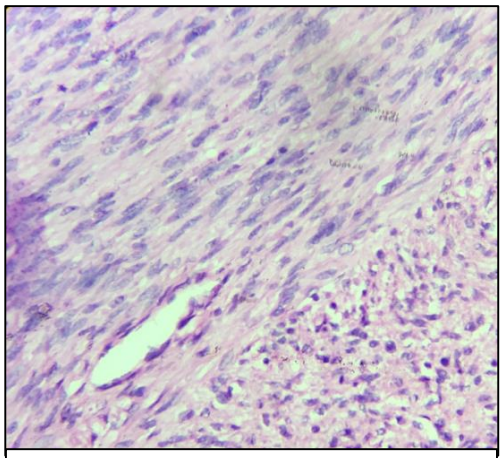
The baby underwent an exploratory laparotomy and right nephrectomy was done. Intraoperatively there were dense adhesions between the tumour and the renal vessels and the sheath of IVC.

The gross pathological examination showed specimen of kidney with a well defined tumour together weighing 36 gm. Tumour was at one pole of kidney and measured 4x4x3.5cm. Cut section of it was firm, greyish white with whorled appearance. No areas of necrosis or haemorrhage was identified. Renal capsule was intact.

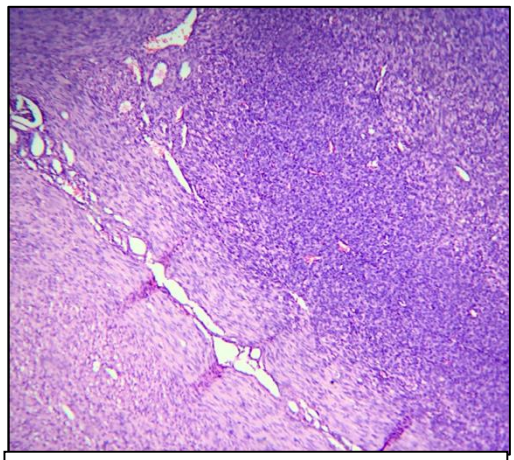
In microscopy, a neoplasm composed of spindle cells arranged in interlacing fascicles was seen which dissected and entrapped islands of normal renal parenchyma. Focal areas were highly cellular with diffuse arrangement of cells; round/oval cells with scant to moderate cytoplasm and round/oval vesicular nuclei with fine chromatin and 2-3 mitoses per HPF were noted. Neoplasm involved the renal sinus but cut end of ureter was free of neoplasm. No abnormal mitosis or necrosis noted. A diagnosis of CONGENITAL MESOBLASTIC NEPHROMA-Mixed type (focal highly cellular areas with increased mitosis) Stage II was given.



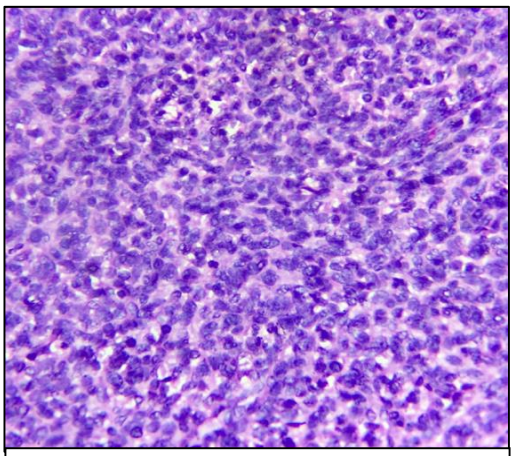
40X. Spindle cells arranged in interlacing



400X. Spindle cells in fascicles



100X. both cellular and classical areas



400X.cellular area with high mitotic count

DISCUSSION:

Neonatal tumours are rare and comprises only 2% of all paediatric malignancies (Powis, M. 2010). In neonates the differential diagnosis for renal neoplasms include CMN, Wilm's tumour, Rhabdoid tumour, Clear cell sarcoma of the kidney and renal cell carcinoma (Al-Turkistani, H. 2008). Congenital Mesoblastic Nephroma is a renal neoplasm most commonly seen during infancy. It is thought to develop from proliferating nephrogenic mesenchyme (Thompson, P., & Chintagumpala, M. 2012). The main symptoms with which patients present are abdominal mass and haematuria. Many cases of CMN are being reported antenatally now. In our case patient presented with abdominal mass. Wilms tumour is a close differential diagnosis of CMN and it is essential to differentiate between the two for proper management. On microscopy the CMN have spindled cell bundles comprised of immature renal stromal cells .CMN tend to infiltrate kidney unlike Wilm's tumour which form a

pseudocapsule. CMN also lack renal blastema and neoplastic metanephric elements.

Histologically CMN are divided into Classical, Mixed and Cellular subtypes. Classic subtype are composed of interlacing fascicles of fibroblastic cells and low mitotic activity. Cellular CMN has a pushing border and are composed of poorly formed fascicles, high mitotic rate and often necrosis. Mixed CMN has features of both classic and cellular subtypes. Cellular Mesoblastic Nephromas are characterized by the translocation (t 12; 15) (p13; q25) which encodes for the ETV6-NTRK3 gene fusion activating the tyrosine kinase growth signalling (Moch, H. 2016).

TREATMENT: Surgery is the mainstay of treatment for CMN i.e Radical Nephrectomy. Local or partial resection is associated with a significantly higher incidence of local recurrence. There is no established role for chemotherapy in the treatment of CMN but for recurrent cellular mesoblastic nephroma chemotherapy appears to have a role. Prognosis is excellent in complete excision. Recurrence is noted in stage III disease, Cellular CMN and age greater than or equal to 3 months (Thompson, P., & Chintagumpala, M. 2012).

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