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Mesoblastic Nephroma with Unusual Presentation: Case Report and Review

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Abstract: ***Congenital mesoblastic nephroma [CMN] is a benign, most frequently occurring mesenchymal renal neoplasm seen in newborns and young infants. We had a 2 - day old neonate with respiratory distress, one episode of hematuria and mass per abdomen. On further evaluation the child was diagnosed as a case of classic congenital mesoblastic nephroma and patient underwent radical nephroureterectomy. The patient is doing well in the post operative period with no recurrence at 6 months follow up.***

Keywords: renal neoplasm in infants, pediatric renal mass, congenital mesoblastic nephroma, neonatal kidney tumor

1. Introduction

CMN is the most frequent type of renal tumor in newborns and young infants, representing about 5% of all renal tumors in pediatric age and approximately 18% of all renal neoplasm during the first 6 months.1 Bolande et al. has distinguished congenital Wilms' tumor from CMN owing to its benign nature.2 Prenatally it is diagnosed as polyhydramnios with renal mass, postnatally usually at 2 months of age.3 The CMN usually presents as an asymptomatic abdominal mass in early infancy. There are few case reports in literature of its presentation with hemorrhagic manifestations with abdominal mass.4 This case report is of a 2 - day old neonate with respiratory distress, one episode of hematuria, abdominal mass and on evaluation was diagnosed as a case of CMN.

2. Case Report

A 2 - day old neonate was evaluated with history of respiratory distress since birth. He was delivered by LSCS in view of fetal bradycardia at 39 weeks. His antenatal scans done at 12th, 24th and 34th week were normal. He had delayed cry at birth which required bag mask ventilation for 30 seconds. Apgar score was 7 at birth and 8 at 5 mins. Exclusive breast feeding was initiated. He had no vomiting, passed stools normally. He had a single episode of hematuria on day two of life. Complete hemogram and coagulation profile were normal. On examination baby had mild tachypnea and a visible mass in right side of abdomen with fullness over right flank. A Mass was size approximately 6X5 cm was palpable in right lumbar region extending up to the midline, non - tender, smooth surface, firm in consistency and ballotable in nature. Ultrasound showed well defined mass lesion of size 53x63 mm in mid and upper pole of right kidney with internal vascularity and echogenicity in renal pelvis, S/o neoplastic etiology. CECT abdomen revealed 5.2x5.3x4.7 cm size mass lesion arising from upper and mid pole of right kidney, with no perirenal infiltration, normal parenchyma was seen in lower pole, right renal vein compressed / thrombosed. Infrarenal IVC showed flow artifact with? partial thrombus which could either be Wilms tumor, or mesoblastic nephroma. Hence FNAC was done which suggested of mesoblastic nephroma. Colour doppler showed good flow in IVC with no evidence of thrombus. Patient was explored with right supraumbilical incision and radical nephroureterectomy was done. Ureter was ligated as low as possible. No suspicious lymph nodes were seen. The renal artery and vein were flush ligated with the inferior vena cava. Total nephrectomy was done and the specimen was sent for HPR. On histopathology examination, the tumor was 5x5x4 cm, infiltrating type composed of spindle shaped cells in sheets and interlacing fascicles. Mild nuclear pleomorphism and infrequent mitosis were noted. The tumor cells were seen infiltrating in between and entrapping the normal parenchyma. Pelvicalyceal system, renal sinuses were involved with no capsular, ureterovascular and lymphovascular invasion. Resected margin of perinephric fat was free of tumor. Overall, the features were suggestive of Classic congenital mesoblastic nephroma. Postoperative duration was uneventful with no signs of recurrence at 6 months of follow - up.

3. Discussion

Solid renal neoplasms are uncommon in neonatal age. CMN is the most frequent renal tumor seen in neonates and early infancy. The tumor which was previously termed as fetal renal hamartoma, leiomyomatous renal hamartoma was coined as "congenital mesoblastic nephroma" by Bolande et al. in 1967[2]. Typically, CMN presents as an asymptomatic abdominal mass at 2 months of life with an equal male and female preponderance. These lesions can also be diagnosed on antenatal ultrasound scans usually in third trimester on careful evaluation.

These tumors are associated with polyhydramnios (40 - 70%) on prenatal evaluation.5 A minority of patients may present with hematuria (11%), hypertension (19%), vomiting, hypercalcemia, jaundice, dehydration, azotemia, and electrolyte disturbances postnatally.

Kamaraj et al stated that neonates presenting with hematuria as the primary complaint should be extensively evaluated and should undergo serial imaging to rule out any occult renal neoplasms. Hematuria in this case was transient in nature, single episode which resolved spontaneously without any intervention.4Hypertension in CMN has been attributed to hyperreninemia, which could be primary - secreted by the tumor cells or secondary due to compression of the renal parenchyma by the tumor inducing ischemia which is more

common in the leiomyomatous type.7 However the blood pressure was normal in our case.

The CMN has been classified histologically into Classic (65%), Cellular (25%) and Mixed types (10%) depending of the nature and arrangement of cells. The classic variant has been described as a solid tumor with fusiform spindle cells, with low rates of mitosis and no necrosis. The cellular variant has sarcomatous appearance with high cellularity, increased rates of mitosis, presence of necrosis, hemorrhage and reduced cytoplasm with ovoid or fusiform spindle cells. The Mixed - type variant has features of both the above variants in varied proportions[8].

Genetic factor i. e. translocation t (12; 15) which results in fusion of the ETV6 gene on chromosome 12p13 and NTRK3 gene on chromosome 15p15 has been described exclusively for the cellular - type vatriant.9, [10] Genetic assessment was not done in this case as our patient had features of classic variant.

CMN generally carries an excellent prognosis with a disease - free survival of 94% and overall survival (OS) of 95%.

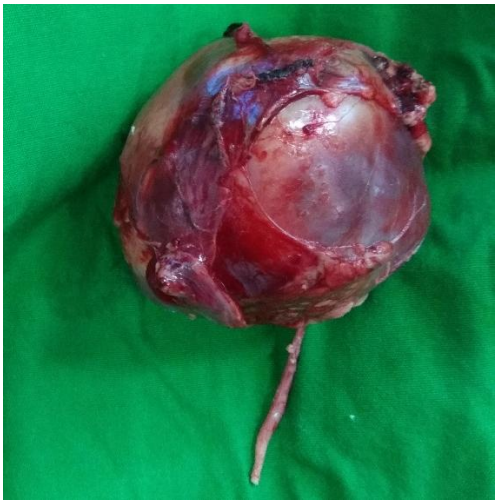
Radiological features although nonspecific, should be used in conjunction with age, presenting symptoms and clinical findings to make the diagnosis. Factors such as adequacy of resection, age of presentation, cellular variant of CMN.[4, 9, 10.] have to be kept in mind while treating a case of CMN as these tumors tend to grow into the hilar region compressing the vessels, could spread to local peritoneal tissue or distantly metastasize to lung, liver as described by various studies, recurrence which has been documented.

Nephrectomy is the primary modality of treatment. Kalidasan et al suggested that surgical excision which could either be a total nephrectomy or partial nephrectomy if negative margins were achievable.1[1]However in this case the tumor was seen involving the upper pole and extending to central location which made it unsuitable for partial nephrectomy. The role of chemotherapy remains controversial. Nevertheless, neoadjuvant chemotherapy with vincristine and actinomycin have been tried, results of which are equivocal. Role of adjuvant chemotherapy in treatment is doubtful and should be used in case of recurrence or metastatic diseases. Radiotherapy is not routinely used owing to the delayed effects of radiotherapy and be cautiously used in cases of aggressive tumors not responding to chemotherapy with caution.4, [9, 10.]

4. Conclusion

CMN though rare should be considered as a differential diagnosis for renal mass occurring in infancy.

The presence of an abnormality in the contralateral kidney, congenital Wilms should be considered. Both the classic and cellular variants of CMN can be treated with nephrectomy alone if wide surgical margins are obtained, especially if the tumor is diagnosed and treated before 6 months of age. For patients with stage III cellular CMN chemotherapy is an option though the efficacy is uncertain, not recommended in stage I - II cellular CMN and generally has good prognosis. A close follow - up is necessary for at least 1 year as there are chance of recurrence.



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