

CASE REPORT

Recurrent Congenital Mesoblastic Nephroma: A Case Report

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ABSTRACT

Congenital mesoblastic nephroma (CMN) is the most common tumor of kidney in early infancy. We present here a case of antenatally detected right renal mass that was excised and reported cellular CMN. He developed recurrence while on adjuvant chemotherapy. We emphasize that the management of cellular CMN, especially adjunct chemotherapy, has not been standardized yet.

Key words: Cellular CMN; Sarcoma; Antenatal

INTRODUCTION

Congenital mesoblastic nephroma (CMN) is mostly a benign tumor of kidney, with nephroureterectomy as a standard of care and usually excellent prognosis. The cellular or atypical CMN is potentially aggressive variant and is known for local recurrence and even distant metastasis. With documented chemosensitivity of CMN, we propose the pendulum should be swung in favor of sarcoma-based chemotherapy.

CASE REPORT

The antenatal scan of a baby at 32 weeks gestation detected right renal mass arising from upper pole with polypoidal and peripheral cystic components (Fig.1). It was associated with retroperitoneal perirenal collection? hemorrhagic. The post-natal computed tomography performed at 3 weeks of age revealed a 6.5 x 5.2 x 4.1 cm right renal mass with solid and cystic components and associated localized peri-renal collection (Fig.2). He underwent right nephro-ureterectomy at day 26 of life elsewhere. Histopathological examination [HPE] revealed cellular atypical mesoblastic nephroma, with foci of tumors showing classical CMN features and definite diffuse infiltration of renal sinus. The tumor cells showed brisk mitosis and expressed Vimentin and cyclin D1; immune-negative for WT-1 and CD34.



Figure 1: Antenatal scan at 32 weeks gestation showing right renal mass (*) with peri-renal collection.

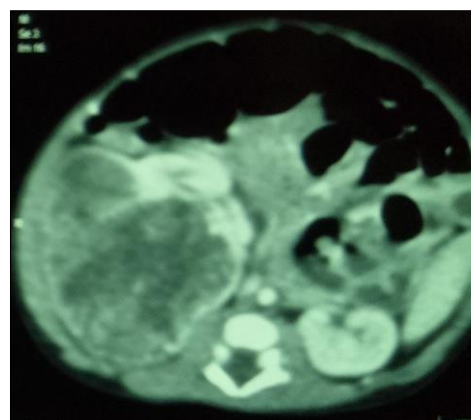


Figure 2: Post-natal CT picture showing right renal mass with localized peri-renal collection.

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He received 13 cycles of Actinomycin-D and vincristine based chemotherapy, starting from 40th post-operative day. At the age of 4.5 months (while on chemotherapy), he developed recurrence that grew to larger than original tumor in coming 6 weeks. CECT of abdomen showed a large complex heterogeneous, predominantly cystic mass in right renal fossa, with multiple hemorrhagic areas (Fig.3). At 6 months age, he was first seen at our center. Exploration revealed a large friable mass infiltrating adrenal gland. The tumor got ruptured intra-operatively; R2 resection was done. HPE showed tumor composed of round by spindled out cells present in sheet, entrapping adrenal gland, clear to eosinophilic cytoplasm, showing brisk mitosis and cystic degeneration, suggestive of cellular CMN (Fig.4).

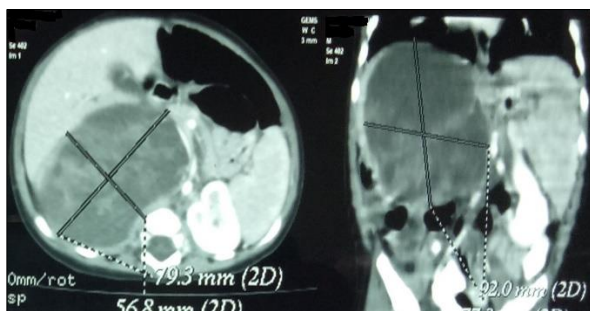


Figure 3: CECT picture showing recurrent right renal fossa mass.

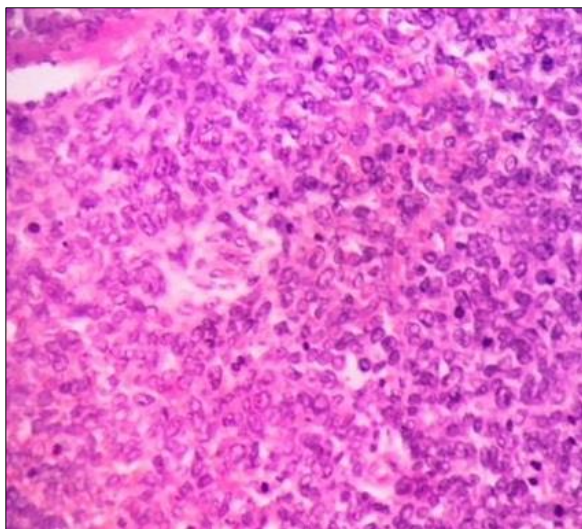


Figure 4: HPE showing round blue tumor cells with clear to eosinophilic cytoplasm, showing brisk mitosis and cystic degeneration, suggestive of cellular CMN.

He received whole abdomen radiation 10.8 Gy over 6 fractions in view of intra-op tumor rupture and 6 cycles of adjuvant chemotherapy (Ifosfamide, Carboplatin and Etoposide- ICE). He has been in close follow up and is disease-free for last 10 months.

DISCUSSION

CMN accounts for 3-10% of childhood renal tumors [1] and commonly present in less than 2 months of age. Two main variants are recognized on histology,

classical and cellular variants. The cellular or atypical CMN is known for local recurrence and even distant metastasis. The genetic aberrations mentioned in CMN include somatic trisomy 11 and t(12;15)(p13;q25), resulting in a fusion of ETV6 and NTRK3.[2,3] Shared histopathology and translocation gene fusion results support the concept of cellular CMN as the renal form of infantile fibrosarcoma (IFS), while classic CMN is equivalent to infantile fibromatosis.[4]

The antenatal diagnosis of CMN and its opponent renal tumors like Wilm's tumor is difficult to conclusively confirm, as many masses show solid and cystic components and even polyhydramnios.[5]

Literature has mainly case reports of CMN and management strategies are based on few published case series. With majority of CMN cases presenting as local disease, surgery is the standard of care. The factors influencing negative outcome for CMN includes stage III, cellular subtype and age older than 3 months.[6] The removal of peri-renal fat is gold standard for CMN, as it often shows infiltrative growth.[7] Stage III CMN represents a dilemma subgroup for adjuvant therapy. A review by Gooskens et al. showed that only 1 case had relapse out of 12 classic and mixed type stage III patients treated without additional chemotherapy.[8] In contrast, 7/12 (58%) stage III cases of the cellular type had relapse who were treated with surgery only, while 4/14 (29%) stage III cellular type cases treated with adjuvant chemotherapy developed a relapse. They proposed that the evidence for additional chemotherapy in stage III cellular CMN is lacking; may be due to small number of cases. England et al. reported 47 cases of CMN treated by surgery alone without any recurrence.[9]

The chemosensitivity of CMN tumor is well documented as evidence by reduction in size or necrosis post neoadjuvant chemotherapy such as Actinomycin-D and vincristine (AV) regimen.[6,9] But the need for neo-adjuvant chemotherapy in CMN may only be indicated in case of large mass or risk of tumor rupture intra-operatively.

The adjuvant chemotherapy for cellular CMN is not standardized. The response data in literature do not corroborate with a specific type of adjuvant chemotherapy.[8] Actinomycin-D based chemotherapy was usually given in 1980s, most of reported chemotherapy later on had combination of chemotherapeutic drugs, usually sarcoma based.[8] Gormley et al. in their study on 7 CMN cases with recurrence showed that Wilm's tumor based chemotherapy (AV) failed to control recurrence in 4 out of 5; out of them 2 later responded to sarcoma based chemotherapy [vincristine, cyclophosphamide and doxorubicin].[10] Role of radiotherapy is controversial. We gave ICE [Ifosfamide, Carboplatin and Etoposide

based] chemotherapy and radiation to whole abdomen in view of intra-operative spillage.

To conclude, AV therapy used for Wilms tumor is not an ideal chemotherapy for cellular CMN and chemotherapy used for sarcomas should be administered to infants with cellular CMN.

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

Author Contributions: All the authors contributed fully in concept, literature review, and drafting of the manuscript and approved the final version of this manuscript.

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