Case Report

MESOBLASTIC NEPHROMA IN CHILDHOOD - DIAGNOSTIC DILEMA : A CASE REPORT

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ABSTRACT

A 11 year old girl presented with gradually enlarging left upper abdominal mass for about 1 year without any urinary or bowel complaints. Hematological, biochemical and urinary examinations reveals no abnormality. Ultrasonography reveals well capsulated mixed echogenic left renal mass. IVU shows a soft tissue mass in left kidney region and poorly excreted, distorted, displaced left kidney which compatible with malrotated crossed ectopic kidney. CT scan shows huge soft tissue mass at left kidney with extension to left real vein which is suggestive of Nephroblastoma. FNAC shows atypical spindle cells - possibility of embryonic/mesenchymal neoplasm (most likely nephroblastoma). Three dose of chemotherapy for Nephroblastoma were given without any response. Finally left radical nephrectomy performed. Histopathology reports a case of Mesoblastic Nephroma (MN). On two years ten months follow-up, there is no evidence of recurrence.

INTRODUCTION

Mesoblastic Nephroma is an uncommon renal tumour of infancy and rarely seen in older- children and adults with highest peak of incidence during the first 3 months of life. Male infants are more frequently affected (male-to-female, 1.8:1). It was previously confused with Wilm's tumor which is rare during first three moths of life. Mesoblastic Nephroma was established as a distinct renal tumor in 1967. It is now classified as a benign tumour of kidney but recurrence and malignant transformation have been reported. There are three subtypes of Mesoblastic Nephroma[1] -

1. Cellular or atypical
2. Classic or leymyomatous
3. Mixed

USG, CT can be done preoperatively to see the extension of disease but cannot give up the exact diagnosis. FNAC may give the diagnosis but there may be diagnostic dilemma. Only histopathological diagnosis is confirmatory. Opinion regarding post-operative Chemotherapy or Radiotherapy is not unanimous. Nephrectomy is the treatment of choice without the need for adjuvant radiation and chemotherapy.

CASE REPORT

A eleven years old girl presented with gradually enlarging painless mass in left upper abdomen for about one year. She had no urinary and bowel complaints. She had no history of significant past illness, no significant family history, no history of consanguinity. She was immunized according to EPI schedule of Bangladesh.

Patient was well-nourished, normothermic, normotensive, nonecteric. She had no cardio-respiratory problem. Her left upper quadrant of abdomen was distended. There was a smooth surfaced, firm, ballotable mass, occupying almost whole of the left upper abdomen crossing the midline. The mass was oval and a bit reiniform in shape. There was no abnormality on auscultation and per rectal examination.

There was no Hematological, Biochemical and Urinary abnormality. USG reveals well capsulated, mixed echogenic left renal mass with tiny calcification. IVU shows normal position and functioning right kidney and a huge soft tissue mass in left kidney region with some contrast within it. Below and right of the mass distorted left kidney noted which is suggestive of crossed ectopia with mal-rotation of left kidney. CT-Scan revealed a huge soft tissue mass (19.4cm x 12.2cm) at left kidney, suggestive of neoplasm (nephroblastoma) with tumor extension/tumor thrombus at left renal vein, and with possible infiltration to left peri-renal lymph nodes.

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FNAC shows spindly cells which is the possibility of embryonic /mesenchymal neoplasm most likely nephroblastoma of left kidney. By taking the diagnosis as left nephroblastoma, 3 dose of new adjuvant chemotherapy for nephroblastoma were given but no response. So we about our diagnosis and we decided to do laparotomy. By left upper transverse generous incision laparotomy performed which reveals a huge soft tissue mass arising from the left kidney with a apparently normal looking rim of kidney tissue in it's lower and medial aspect. The mass occupies almost whole of the left upper abdomen and extended up to right iliac fossa. Left renal vein was free. Left radical nephrectomy performed. There was no spillage of tissue. The mass was weighted 5 kg (Fig -1). Post operative recovery was uneventful.

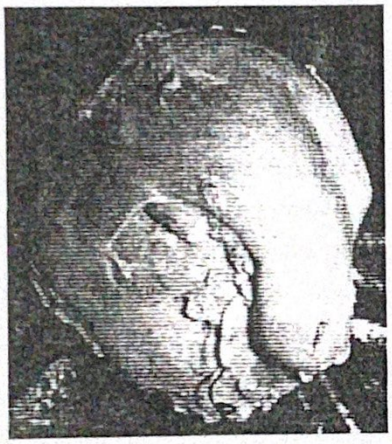

Figure 1 : Specimen of Kidney mass

Fig : Resected Specimen

Histopathology of the mass from home and abroad diagnose the case as Classic type of Mesoblastic Nephroma.

No post-operative chemotherapy given. On three years follow-up there is no evidence of recurrence.

DISCUSSION

Mesoblastic nephroma is a rare benign tumor of kidney with a potential to recurrence and malignant transformation. Mesoblastic nephroma may be associated with Trisomy 11, von Willebrand disease, Beckwith-Wiedemann Syndrome, Multiple Cysts, Polyhydramnios and hypercalcemia, Focal and segmental glomerular sclerosis, but we have no such associated diseases. There is no uniform presentation of Mesoblastic nephroma. Our patient had also no typical clinical and imaging and FNAC findings as it can present with atypical findings. According to some author, it can be diagnosed by clinical, ultrasonographic, CT, MRI and FNAC findings, but often there are diagnostic dilemma with USG, CT and even in FNAC findings. It may present with signs of metastasis but we have no such report. Cytogenetically Mesoblastic Nephroma may be due to Trisomy 11 or may be due to translocation (14;15) (q11;q24). Anatomicopathological finding is the confirmatory diagnosis of Mesoblastic Nephroma. Histopathologically it is characterized by interlacing bundles of spindle Mesenchymal cells. We found no response to chemotherapy as we give chemotherapy for Wilms' tumor but some MNs are chemosensitive specially in case of recurrent or unresectable cellular MN. There are reports of metastasis this tumor to the Brain and liver but we have no such report. Recurrence, even tumor related death have been reported in the literature always related to the atypical form or to the mixed form particularly in patients aged more than three months. Incomplete surgical removal is also one of the cause of recurrence. On three year follow up patient is well without any complications.

CONCLUSION

In case of any renal tumor, when there is pre-operative diagnostic dilemma one can think about Mesoblastic Nephroma. It almost always pursues a benign clinical course but follow up is necessary.

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