Clinical Grand Rounds in Pediatric Oncology

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Case

An 8-day-old female neonate presented with antenatally detected mass in the right kidney.

On Examination

Per abdomen: hard mass with well defined margins palpable in the right hypochondrium and right lumbar region, approximately 6 cm × 10 cm.

Investigations

Usg-abdomen and pelvis with Doppler

Well defined large heterogenous solid lesion of size 5.7 cm × 5.6 cm seen in right kidney medially displacing the Inferior vena cava. IVC upper segment seen but rest of IVC and right renal vein can’t be visualised- probable compression by the mass. Left kidney, liver normal.

CECT- Abdomen: Well-defined heterogeneous enhancing solid mass lesion seen arising from the upper and mid poles of right kidney? Tumor.

Usg guided fnac: Features of Benign spindle cell lesion.

Major surgical profile

PET Scan (Post-operative): No residual tumor, No distant metastasis.

Pre-op management

- Baby admitted in ICU, maintained thermoregulation and vitals monitored
- Administered pre-operative vitamin K and antibiotics and shifted in transport incubator to the operating room for an exploratory laparotomy + precede right nephroureterectomy.

Operative findings

- Right Renal tumour ~8 cm × 5 cm; No enlarged lymph nodes seen
- Ureter identified and mobilized till the pelvic part and ureterectomy done
- Renal vein and artery identified and transfixed
- Right Nephrectomy done preserving right adrenal gland
- Tumor bed margins biopsied and marked with clips

Post-operative management

- To 1st Post-Operative Day (POD)
- Baby was kept in Pediatric Surgery ICU and Nil per Oral
- IV antibiotics & fluids, vitals and abdominal girth charting
- IV Fentanyl infusion for pain relief
- On 2nd POD - Started Breast feeds
- On 5th POD- Child shifted to ward
- On 7th POD- Child discharged from hospital with uneventful recovery

Histopathology Findings

- Well circumscribed tumour tissue with infiltrating margins and composed of cellular growth of spindle cells and interlacing fascicles with papillary , acidophilic cytoplasm and bland spindled nuclei
Figures: 1). 8 cm x 5 cm right renal tumor 2). CECT abdomen showing large renal tumor (red arrow) pushing IVC (green arrow) medially 3). Tumour resected into 4). Cut-section of tumor having uterine fibroid-like appearance.

- There are entrapped glomeruli and tubules in the tumor tissue
- Focal areas show cellular proliferation with increased mitotic activity
  - There is no invasion into renal pelvis or post renal fat or lymphovascular invasion
  - All margins are free from tumor
- Based on operative and histopathology findings it was classified as right congenital mixed mesoblastic nephroma (classical + cellular)

**Adjuvant Chemotherapy**

In lieu of mixed variant on histopathology with potential for recurrence, PET scan was done which ruled out any other distant metastasis and baby underwent 8 weeks of Vincristine + Actinomycin-D chemotherapy in our ward which she tolerated very well.

**Present status**

Baby is 5 years old now, healthy and disease free.

**Congenital Mesoblastic Nephroma**

Congenital Mesoblastic nephroma is a rare neonatal/pediatric renal tumor that is usually found before birth by USG or within the first 3 months of life. It is similar in gross and histologic appearance to a uterine leiomyoma with spindled cell bundles, but composed of immature renal stromal cells. The tumor lacks renal blastema and neoplastic metanephric elements, thereby differentiating it from Wilm’s tumor. It can be detected antenatally especially with judicious use of ultrasonography. There are two pathologic variants: classic CMN and atypical or cellular CMN. The classic form is characterized by rare mitoses and absence of necrosis. Atypical or cellular CMN is characterized by a high mitotic index, hypercellularity, necrosis, hemorrhage, and invasion of adjacent structures warranting use of adjuvant chemotherapy.

**Therapeutic management**

Upfront surgical excision + Chemotherapy + Radiotherapy

**Points to Remember**

- Cancer can occur in children – even in newborn babies!!
- Cancers in babies can sometimes be detected even before birth by antenatal ultrasonography
- Cancers in children are treatable and when detected early have very good outcomes
- Paediatric cancers do not cause associated symptoms like weight loss, weakness except in late stages - so do not wait for symptoms
  - Do not waste time & seek urgent pediatric specialist care.