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WILM'S TUMOR (A SERIES OF 3 CASES)

Paediatric Surgery

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ABSTRACT

Globally, Wilm's tumour accounts 6% of all paediatric malignancies with overall survival rates over 90%.The Children Oncology Group recommends to go ahead with a radical nephrectomy followed by the need of radiation and chemo therapy depending on the staging of the disease. **Aim:** To study overall outcomes of patients with Wilm's tumour. **Methodology:** All the patients were <18 years, of either gender with symptoms of lump and distention of abdomen, were evaluated with USG and CECT abdomen+pelvis, clinicoradiologically diagnosed as Wilm's tumor. **Presentation Of Cases:** Overall 3 cases were presented in this case series. **First case** of this study illustrates the diagnosis of Wilm's Tumor (stageII) in a patient who was posted for right sided nephrectomy. Patient was discharged on POD14. Patient received 2 drug adjuvant chemotherapy regimen of Inj vincristine and actinomycin-D post nephrectomy and is now healthy without any complication. **Second case** described a 1 year and 6months old hypertensive male with distended abdomen and palpable lump in abdomen since 3 months, was diagnosed with left nephroblastoma with Grade II Hypertensive Retinopathy who underwent left total nephrectomy and received 14 cycles of inj vincristine post nephrectomy. The patient described in **third case**, was a k/c/o left nephroblastoma, received neoadjuvant chemotherapy 12 cycles of Injection Vincristine. And then underwent left total nephrectomy. **Conclusion:** There are specific patient populations that continue to have poor overall survivals, a high risk for late effects, or the potential for significant treatment reductions despite the excellent overall outcomes for children with renal tumors. Understanding and treating patients with nephrogenic pathology require additional research.

KEYWORDS

INTRODUCTION:

The most frequent renal tumor in children (typically younger than the age of 5) is Wilms tumor (nephroblastoma), which has an overall survival rate of over 90%. [1].For a number of reasons, this tumor is being researched. The first is the need to reduce long-term side effects brought on by current treatment modalities, and the second is the need to find more potent treatments for patients with high-risk histological subtypes and those who have relapsed.[2].It makes up 6% of all pediatric malignancies worldwide, with bilateral cases of Wilms' tumor accounting for 5-7% of all cases. [3]

As of now, there is no evidence-based agreement on Wilm's tumor management. In order to assess for venous thrombosis and tumor extension during nephrectomy, it is necessary to palpate the renal vein and sample sentinel lymph nodes. [4]

PRESENTATION OF CASES:

CASE NO. 1-

A 4 months old male patient presented with a lump and distention of abdomen since 20 days. On per abdomen examination a lump of approx.size 4x3cm is palpable in right subcostal region extending up to lumbar region, hard in consistency, non-tender. Radiological investigations revealed, right kidney is enlarged (9.5x5.5cm) and impaired with tumour involving complete parenchyma, s/o Wilm'stumour on ultrasonography of abdomen &pelvis;On CECT abdomen + pelvis a large well defined solid hypodense lesion arising from anterior aspect of kidney extending from superior to inferior pole represents neoplastic lesion probably Wilms'stumour. All blood investigations were within normal limits. Patient was diagnosed on the basis of clinical and radiological findings with Wilms'stumour. Patient was posted for right sided nephrectomy and specimen was sent for histopathological examination, which revealed Wilm's tumour with capsular invasion of right kidney, stage II. Patient received 2 drug adjuvant chemotherapy regimens of inj vincristine and actinomycin-D.

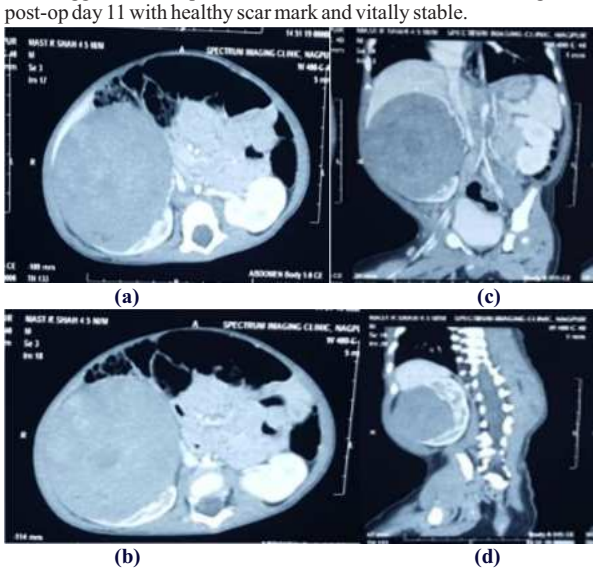
CASE NO 2-

A 1 and half yearhypertensive male patientcame to the OPD with distended abdomen and palpable lump in abdomen since 3 months, which was insidious in onset and gradually progressive in nature. Both mother and child (patient) have no history of any congenital anomalies and no significant family history. On per abdomen examination a lump of size 15x15cm (approx.) was palpable in left side of abdomen extending from left subcostal region to left iliac region. On fundus examination patient had Grade II Hypertensive Retinopathy. Radiological examination was done, ultrasonography of abdomen& pelvisrevealed a well-defined heterogenous lesion of size 15x9.5x15 occupying the entire abdomen likely arising from the left lumbar region and displacing the adjacent organs, lesion crossing the midline and shows multiple cystic space within and mild internal vascularity within. CECT Abdomen + Pelvis revealed a large, well defined, heterogenous, solid, cystic lesion measuring 15.6x13.3x14.6cm in AP, transverse and CC dimensions extending from T7 to S1 vertebrae is noted arising likely from the upper pole and interpole region of left kidney was seen, most likely represents Nephroblastoma (Wilms's Tumor) and right kidney normal in location, shape, and adequate excretion of contrast.All other blood investigations were within normal limit.Patient was diagnosed with Left Nephroblastomaon the basis of clinical and radiological findings and was posted for left sided total nephrectomy was made. Patient underwent total nephrectomy and specimen was sent for Histopathological Examination. Gross Examination reports were suggestive of a single, large, globular tumour mass with bosselated external surface of weight 1.25kg seen. On microscopic examination, undifferentiated blastemal cells and cells differentiating mostly in epithelial lineage and scanty stromal elements with few areas of necrosis seen. Histological feature was consistent with Wilms'stumour. Patient received 1st Chemotherapy cycle of Inj. Vincristine 0.5mg on POD-13 and was discharged on post-op day 14. Patient received 14 more cycles of Injection Vincristine0.5mg with 1 week of interval in between in follow-up.

CASE NO 3-

A 13 years old female patient k/c/o Left Nephroblastoma presented to the OPD withpalpable lump in abdomen since 6months, which was insidious in onset and gradually progressive in nature. Both mother and child had no significant family history.On per abdomen examination a lump of size 16x12 cm (approx.) was palpable in left side of abdomen extending from left hypochondrium to left iliac region.Patient had completed 12 cycles of Injection Vincristine as the mass was clinically inoperable as it was crossing the midline . On radiological examination post chemotherapy, CECT Abdomen + Pelvis revealed left kidney is completely replaced by a large relatively

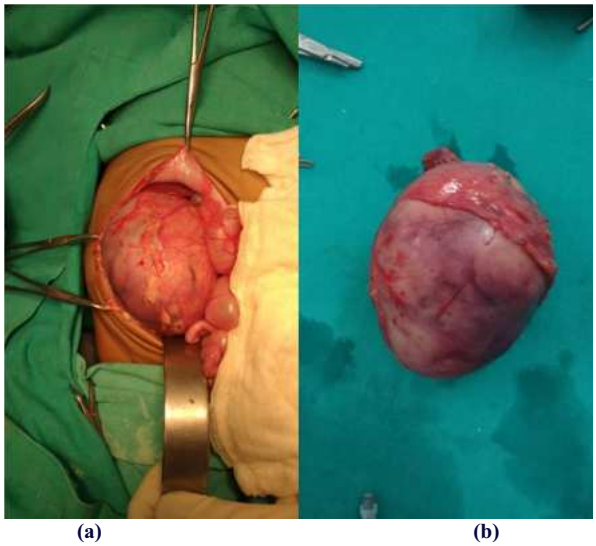
well defined solid cystic soft tissue attenuation lesion of size 7.7x12.5x16.5cm with thickness measuring 3.3mm, likely suggestive of neoplastic aetiology in left kidney. Blood investigationswere within normal limit. Patient underwent left Total Nephrectomy. Specimen was sent for Histopathological Examination. Histological features were suggestive of nephroblastoma tumor. Patient was discharged on post-op day 11 with healthy scar mark and vitally stable.



(a) (c)

(b) (d)

Figure1. (a,b,c,d) CT scan of 4month old patient showing a large well defined solid hypodense lesion arising from anterior aspect of kidney extending from superior to inferior pole.



(a) (b)

Figure2. (a) Intra-operative tumor ,(b) Post-Nephrectomy Specimen

Figure 4. Post Operative Surgical Scar

The majority of Wilm's tumor patients have no symptoms, but some might have an abdominal mass, pain in the abdomen, weight loss, anorexia, haematuria, or hypertension. [5].As a tumor of the renal parenchyma, Wilm's tumor typically grows towards the renal hilum. [6]. The complete pathologic evaluation, accurate nodal staging, and clearance of all local disease are the objectives of primary surgery for unilateral WT. Unilateral radical nephroureterectomy with lymph node sampling is the suggested course of action, and this recommendation is backed up by a number of cooperative trials. Through a transabdominal or thoracoabdominal incision. [7].

First case of this study illustrates the diagnosis of Wilm's Tumor (stageII) in a patient who was posted for right sided nephrectomy. Patient was discharged on POD14. Patient received 2 drug adjuvant chemotherapy regimen of inj vincristine and actinomycin-D post nephrectomy and is now healthy without any complication.

Second case described a 1 year and 6months old hypertensive male with distended abdomen and palpable lump in abdomen since 3 months, was diagnosed with left nephroblastoma with Grade II Hypertensive Retinopathy who underwent left total nephrectomy and received 14 cycles of inj vincristine post nephrectomy.

The patient described in **third case**, was a k/c/o left nephroblastoma, received neoadjuvant chemotherapy 12 cycles of Injection Vincristine. And then underwent left total nephrectomy.

DISCUSSION

Renal tumors account for 6.3% of cancer diagnoses for children younger than 15 years of age, with a reported incidence of 7.9 per million. Including adolescents younger than 20 years of age, this drops slightly to 4.4% of cancer diagnoses, with an incidence of 6.2 per million.[1] Renal tumors include Wilms' tumor (WT) (also referred to as nephroblastoma or renal embryoma), renal cell carcinoma (RCC), clear cell sarcoma of the kidney (CCSK), rhabdoid tumor of the kidney (RTK), congenital mesoblastic nephroma, cystic renal tumor, and angiomyolipoma.[2,3]WT is by far the most common, accounting for approximately 91% of all renal tumors in childhood. The treatment strategy for children with renal tumors evolved in conjunction with the definition of these pathologic subtypes. Treatment is based on traditional risk factors, stage and histology, and, more recently, on genetic markers. The goal of "risk-based management" is to maintain excellent outcomes but at the same time spare children with low-risk tumors intensive chemotherapy and radiation, with their long-term side-effects, and to intensify therapy for children with high-risk tumors in an effort to increase their survival. WT is the most common primary malignant renal tumor of childhood and comprises 6% of all pediatric tumors.[6,8]Outcomes for children with WT improved dramatically over the last 50 years, with long-term survival in both North American and European trials approaching 85% (Fig. 30-1). Survival rates for many of the low-stage tumors are 95% to 99%.[5,6]

TABLE 155-5 Staging System of the Children's Oncology Group

STAGE	
I	Tumor confined to the kidney and completely resected. The renal capsule is intact and the tumor was not ruptured before removal. No renal sinus extension. There is no residual tumor.
II	Extracapsular penetration, but tumor is completely resected. Renal sinus extension or extrarenal vessels may contain tumor thrombus or be infiltrated by tumor.
III	Residual nonhematogenous tumor confined to the abdomen: lymph node involvement, any tumor spillage, peritoneal implants, tumor beyond surgical margin either grossly or microscopically, or tumor not completely removed.
IV	Hematogenous metastases to lung, liver, bone, brain, and so on.
V	Bilateral renal involvement at diagnosis.

Current treatment protocols for children with WT were developed through a series of multidisciplinary cooperative group trials in both North America and Europe by the Children's Oncology Group (COG), formerly the National Wilms' Tumor Study Group (NWTSG), and the

Socie´te´ Internationale d'Oncologie Pe´diatrique (SIOP). There are differences between the approaches of these two groups that affect staging and risk classification that are critical to understand when considering outcomes that will be discussed later in the clinical presentation Most children with WT present with an asymptomatic abdominal mass, often discovered by either a parent or pediatrician. Nonpalpable tumors are typically discovered by ultrasonography during evaluation for abdominal pain. Gross hematuria has been reported in 18.2% of patients and microscopic hematuria in 24.4%. Ten percent of children with WT have coagulopathy, and 20% to 25% present with hypertension because of activation of the renin-angiotensin system. Fever, anorexia, and weight loss occur in 10%. Extension of tumor thrombus into the renal vein can obstruct the spermatic vein and result in a left varicocele and, in rare cases, tumorextension into the atrium may produce cardiac malfunction. Tumor rupture and hemorrhage are also infrequent events that can present as an acute abdomen. After an abdominal mass is identified, radiographic imaging is performed to determine the anatomic location and extent of the mass. Ultrasonography (US) is a good screening examination of a mass to determine its site of origin and to assess for possible intravascular or ureteral extension. About 4% of WT present with inferior vena cava (IVC) or atrial involvement and 11% with renal vein involvement.[6,8] Embolization of a caval thrombus to the pulmonary artery can be lethal, and the presence of a thrombus must be identified preoperatively to prevent this occurrence. US is a sensitive technique to identify vascular extension. .A computed tomography (CT) scan of the abdomen will confirm the renal origin of the mass and determine whether there are bilateral tumors. Early generations of CT scans missed 7% to 10% .MRI is currently being evaluated as a method to help distinguish nephrogenic rests from WT and may be the preferred method to follow children with bilateral WT after resection. The common sites of metastatic spread are the lungs and the liver. Therefore, in addition to abdominal imaging, pulmonary imaging must be performed. WT are embryonal tumors containing components seen in normal developing kidneys. The classic WT consists of three elements: blastemal, stromal, and epithelial tubules. Tumors contain various proportions of each of these elements. Triphasic patterns containing blastemal, stromal, and epithelial cell types are the most characteristic, but biphasic and monophasic lesions occur. Less frequently, abnormal mucinous or squamous epithelium, skeletal muscle, cartilage, osteoid, or fat are found in WT.The COG/NWTS and SIOP staging systems are fundamentally different. In COG/NWTS protocols, initial surgical resection is recommended in most cases. Thus for unilateral tumors, the pathology of the tumor is established prior to administration of chemotherapy or radiotherapy. In contrast, SIOP protocols generally recommend chemotherapy followed by nephrectomy, and surgicopathologic staging is assessed at that time. Surgical therapy is a primary component in the multidisciplinary treatment of WTor other neoplastic renal lesions. Irrespective of whether surgery is performed as a primary therapy or in a delayed fashion after chemotherapy, there are a number of fundamental tasks that are required of the surgeon. These are (1) safe resection ofthetumor, (2) accurate staging ofthetumor, (3) avoidance of complications that will “upstage the tumor” (rupture or unnecessary biopsy), and (4) accurate documentation of operative findings and details of the procedure in the operative note. Intraoperative events that negatively affect patient survival include tumor spill, failure to biopsy lymph nodes, incomplete tumor removal, failure to assess for extrarenal tumor extension and surgical complication.

On the COG renal tumor protocols, treatment is dependent on surgical and pathologic staging after immediate nephrectomy. There are, however, some situations wherein preoperative chemotherapy is recommended. These include children for whom renal-sparing surgery is planned (Blute et al, 1987), tumors inoperable at surgical exploration (Ritchey et al, 1994), and tumor extension into the IVC above the hepatic veins (Ritchey et al, 1993b; Shamberger et al, 2001; Szavay et al, 2004). The last two conditions are associated with an increased risk for surgical complications if primary nephrectomy is performed (Ritchey et al 1992).

CONCLUSION:

Even though children with renal tumors generally have excellent outcomes, there are specific patient populations that continue to have low overall survival rates, a high risk for late effects, or a chance of significant treatment reduction. In upcoming studies of renal tumors, surgical objectives include lowering lymph node sampling failure. To comprehend and treat patients with nephrogenic pathology, more research is required. Future studies will continue to assess the role that tumor biology plays in risk stratification in addition to tumor stage and pathology. [8].

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