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Review article

Wilms tumour-An overview

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ABSTRACT
Wilms tumor (nephroblastoma), an embryonal malignancy of the kidney, is the most common renal tumor of childhood. Wilms tumor usually presents as an abdominal mass in an otherwise apparently healthy child. Wilms tumor has the potential for both local and distant spread. Approximately 5%-10% of children with Wilms tumor have bilateral or multicentric tumors. The average age at presentation is 42-47 months for children with unilateral Wilms tumor and 30-33 months for those with bilateral Wilms tumor.Treatment options include surgery,chemotherapy and radiation therapy.wilm's tumour is almost curable by the Multidisciplinary approach with a good team work of surgeon,oncologist and radiotherapist.
Keywords : wilms tumour,children, nephroblastoma.

Introduction

Wilms tumor , or nephroblastoma is cancer of the kidneys that typically occurs in children, rarely in adults[1]. It is named after Dr.Max Wilms, the German surgeon (1867–1918) who first described it. [2,3]

Epidemiology

- Wilms' tumour is the second most common intraabdominal cancer of childhood and the fifth most common paediatric malignancy overall (approximately 6% of all paediatric cancers).
- Only 3% of Wilms' tumours are reported in adults. Most adult patients are diagnosed unexpectedly following nephrectomy for presumed renal cell carcinoma.
- In 5-10% of patients, both kidneys are affected either at the same time (synchronous bilateral Wilms' tumour) or one after the other (metachronous bilateral Wilms' tumour).

The disease occurs in about 1 out of 200,000 to 250,000 children. It usually strikes when a child is about 3 years old. It rarely develops after age 8.

Etiology

Exact etiology of the wilms tumour is not known .Only a few risk factors for kidney tumors are known for sure. Children with some genetic syndromes and abnormalities present at birth are more likely to develop Wilms tumor than other children. The conditions are aniridia (absence of the iris, the coloured part of the eye), abnormalities of the urinary tract, hemihypertrophy (enlargement of one side of the body), Beckwith-Wiedemann syndrome, Perlman syndrome, Denys-Drash syndrome and Simpson-Golabi-Behmel syndrome. [12-17]

RISK FACTORS

Factors that may increase the risk of Wilms' tumor include:

- Female gender
- Black race
- Family history of Wilms' tumor.

PATHOGENESIS

once study suggest that the the insulin−like growth factor−II (IGF2) and H19 genes are imprinted in human, with expression of the paternal IGF2 and maternal H19alleles. IGF2 undergoes loss of

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imprinting (LOI) in most Wilms' tumours (WT). They conclude that the (i) LOI of IGF2 is associated with a 80−fold down regulation of H19 expression; (ii) these changes are associated with alterations in parental−origin−specific, tissue−independent sites of DNA methylation in the H19 promoter; and (iii) loss of heterozygosity is also associated with loss of H19 expression. Thus, imprinting of a large domain of the maternal chromosome results in a reversal to a paternal epigenotype. These data also suggest an epigenetic mechanism for inactivation of H19 as a tumour suppressor gene.

Current models of Wilms tumor development propose that a genetic mutation predisposes to nephrogenic rests, benign foci of embryonal kidney cells that persist abnormally into postnatal life. Nephrogenic rests are found in approximately 1% of newborn kidneys and usually regress or differentiate by early childhood . Some nephrogenic rests persist into childhood. These rests are considered to be Wilms tumor precursors; nephrogenic rests that sustain additional mutations transform into a Wilms tumor .

Nephrogenic rests are characterized as intralobar or perilobar:

- Intralobar rests are usually solitary and randomly distributed throughout the kidney, although they tend to be situated centrally within the renal lobe. Intralobar rests are associated with two syndromes related to WT1 mutations: WAGR (Wilms tumor-aniridia-genital anomalies-retardation) syndrome (see Aniridia) and Denys-Drash syndrome (DDS) [Breslow et al 2006].

- Perilobar rests tend to be located at the periphery of the kidney and are often multiple. Perilobar rests are associated with Beckwith-Wiedemann syndrome (BWS) and hemi hyperplasia.

The association between type of nephrogenic rest and predisposition syndrome is not absolute .

The term nephroblastomatosis is used to describe the presence of multiple nephrogenic rests. Nephroblastomatosis may be manifest as a diffuse overgrowth of rests (producing a rim that enlarges the kidney) or as multiple distinct rests . It is sometimes challenging to distinguish nephrogenic rests from Wilms tumors, even with biopsies. Although nephrogenic rests are considered benign, chemotherapy has been advocated if the rests are growing or if a child becomes symptomatic. Some evidence indicates that chemotherapy may decrease the risk for subsequent Wilms tumor development in children with nephrogenic rests .

Table 1. Association of Nephrogenic Rests with Wilms Tumor Predisposition Syndromes and Congenital Anomalies

Clinical Phenotype	+ ILNR − PLNR	− ILNR + PLNR	+ ILNR + PLNR	− ILNR − PLNR
Denys-Drash	59%	0%	4%	37%
WAGR	73%	4%	4%	18%
Beckwith-Wiedemann syndrome	18%	35%	27%	20%
Male GU anomalies	43%	9%	5%	44%

ILNR = intralobar nephrogenic rests
PLNR = perilobar nephrogenic rests
WAGR = Wilms tumor-aniridia-genital anomalies-retardation
GU = genitourinary

PATHOLOGY

Most nephroblastomas are unilateral, being bilateral in less than 5% of cases. They tend to be encapsulated and vacularized tumors that do not cross the midline of the abdomen. In cases of metastasis it is usually to the lung. A rupture of Wilms tumor put the patient at risk of hemorrhage and peritoneal dissemination of the tumor. In such cases, surgical intervention by a surgeon who is experienced in the removal of such a fragile tumor is imperative.

Pathologically, a triphasic nephroblastoma comprises three elements:

- Blastema
- Mesenchyme
- Epithelium

Wilms' tumor is a malignant tumor containing metanephric blastema, stromal and epithelial

derivatives. Characteristic is the presence of abortive tubules and glomeruli surrounded by a spindled cell stroma. The stroma may include striated muscle, cartilage, bone, fat tissue, fibrous tissue. The tumor is compressing the normal kidney parenchyma.

The mesenchymal component may include cells showing rhabdomyoid differentiation. The rhabdomyoid component may itself show features of malignancy (rhabdomyosarcomatous Wilms).

Wilms' tumors may be separated into 2 prognostic groups based on pathologic characteristics:

- "Favorable"(95%) - Contains well developed components mentioned above
- "Anaplastic"(5%) - Contains diffuse anaplasia (poorly developed cells)

➢ **Favorable versus Unfavorable Histology in Wilms Tumor:**

	Favorable Histology (FH)	**Unfavorable Histology (UH)**
Features	• Prominent tubular differentiation • No anaplasia • No sarcomatous elements	Extreme anaplasia 3 cytological abnormalities: • hyperdiploid mitotic figures • nuclear enlargement (3x or more) • hyperchromasia Anaplasia may be diffuse or focal; even in one or two foci is enough to impart a markedly worse prognosis These changes are associated with high rates of relapse and death
Proportion of tumors	• 85-88% of WT cases	• 10% of WT cases
	• Outlook with current therapy is good	• 60% of deaths due to Wilms tumor have this histology

About 2% of Wilms tumors have ureteral involvement. Presence of gross hematuria, nonfunctioning kidney, or hydronephrosis suggests the tumor may extend into the ureter and cystoscopy is recommended

PRESENTATIONS

Symptoms

The most common manifestation of Wilms tumor is an asymptomatic abdominal mass which occurs in 80% of children at presentation. Abdominal pain or hematuria occurs in 25%. Urinary tract infection and varicocele are less common findings than these. Hypertension, gross hematuria, and fever are observed in 5-30% of patients. A few patients with hemorrhage into their tumor may present with hypotension, anemia, and fever. Rare patients with advanced disease may present with respiratory symptoms related to lung metastases. [12]

DIAGNOSIS

Physical examination

Physical Examination often reveals a palpable abdominal mass. The abdominal mass should be carefully examined. Palpating a mass too vigorously could lead to the rupture of a large tumor into the peritoneal cavity.

Commonly performed Tests in the diagnosis of wilms tumour include:

- Abdominal ultrasound
- Abdominal x-ray
- BUN
- Chest x-ray
- Complete blood count (may show anemia)
- Creatinine
- Creatinine clearance
- CT scan of the abdomen
- Intravenous pyelogram
- Urinalysis

Other tests may be required to determine if the tumor has spread.

Imaging

The workup of a child with suspected Wilms tumor begins with appropriate diagnostic imaging studies to define the extent of disease and to help plan the surgical intervention.

- Ultrasonography is the recommended first-line test for children suspected of having Wilms tumor.
- Computed tomography (CT)
- Magnetic resonance imaging (MRI) is not a routine component of the evaluation of Wilms tumor, although MRI is being used with increasing frequency because MRI may facilitate the distinction between Wilms tumor and nephrogenic rests.

- Positron emission tomography : PET may play a role in the detection of occult metastatic sites at recurrence.

Surgical resection or biopsy

Although imaging studies may suggest a diagnosis of Wilms tumor, the definitive diagnosis can be made only on histologic assessment of the tumor.

- The COG recommends performing nephrectomy/tumor resection and regional lymph node sampling before chemotherapy to obtain the most accurate staging information. If the tumor is deemed unresectable, a biopsy is recommended to confirm the diagnosis .
- The International Society of Pediatric Oncology (SIOP) recommends administering preoperative chemotherapy to all individuals (with or without a biopsy, depending on the individual's age) to shrink the tumor with the aim of facilitating surgery

Differential Diagnosis of Wilms Tumor

Conditions to be considered in the differential diagnosis of Wilms tumor include the following:

- Mesoblastic nephroma - Most common renal tumor in the first month of life
- Renal cell carcinoma
- Clear cell sarcoma of the kidney
- Rhabdoid tumor of the kidney
- Nonmalignant mass
- Hydronephrosis
- Multicystic kidney disease
- Renal cyst
- Renal thrombosis
- Dysplastic kidney
- Renal hemorrhage
- renal sarcoma
- renal carbuncles
- Neuroblastoma

STAGING

Staging is determined by combination of imaging studies and pathology findings if the tumor is operable. Stage and histopathology are the most important determinants of outcome in children with WT. Treatment strategy is determined by the stage.

There are currently two staging systems available reflecting treatment differences. The current system used by the COG reflects staging at primary surgery. Alternatively, the system used by the International Society of Paediatric Oncology (SIOP) is performed after preoperative chemotherapy.

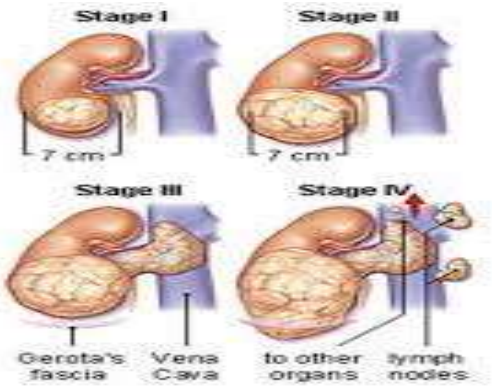
Tumors with favorable histology are more likely to be of lower stages versus anaplastic tumors, which are twice as likely to have Stage IV disease .

Table 2 Children's Oncology Group Wilms' Tumor Staging System

Stage I Tumor confined to kidney and completely resected; no capsular breach, tumor spillage or renal sinus extension
Stage II Extracapsular penetration (including iatrogenic via biopsy prior to resection) or renal sinus extension with vascular involvement; complete resection with negative margins and no lymph node involvement
Stage III Non-hematogenous spread beyond the kidney (abdominal lymph nodes, transected renal vein, IVC tumor thrombus); macroscopic/microscopic residual tumor after resection; peritoneal spillage during resection
Stage IV Hematogenous metastases (lung, liver, bone, brain) or extra-abdominal lymph node spread
Stage V Bilateral renal involvement at diagnosis

Table 3 International Society of Paediatric Oncology Staging System

Stage I Tumor limited to kidney; fibrous pseudocapsule surrounds tumor if outside of contours of kidney; clear resection margins; no renal sinus vessel Involvement
Stage II Tumor extends beyond kidney into perirenal fat, renal sinus, adjacent organs or IVC; complete resection with clear margins
Stage III Incomplete excision of tumor; positive abdominopelvic lymph nodes, tumor penetration through peritoneal surface, tumor thrombi at vascular resection margins
Stage IV Hematogenous metastases; extra-abdominopelvic lymph node metastases
Stage V Bilateral tumors at diagnosis



TREATMENT

Wilms' tumor is the most common malignant tumors of the kidney in children. The treatment of Wilms' tumor can be considered as the paradigm for multimodal treatment of malignant solid tumors in childhood. Progress has occurred from the times when this tumor was universally fatal to this era when more than 85% of the patients can be completely cured with localized disease and over 70% for metastatic disease.

Major research and randomized controlled trials performed by several co-operative groups have made the future of Wilms' tumor patients very bright. The two major groups which have tremendous contributions in the management of Wilms' tumor are National Wilms' Tumor Study (NWTS) and the Societe Internationale D'oncologie Pediatrique (SIOP).

Surgery maintains an important role in treatment, although the improved prognosis for this malignancy during the 20th century is attributed primarily to advances in chemotherapy.Overall survival rates reach 90% with current

treatment regimens. COG and SIOP treatment protocols are now focusing not only on maximizing cure, but also minimizing treatment side effects and associated morbidity. [22-29]

National Wilms Tumor Study Group (NWTS)

A major collaborative effort was the **N**ational **W**ilms **T**umor **S**tudy **G**roup founded in 1969. This cancer research cooperative group set up studies that involved many different treatment centers principally in North America.

NWTS was responsible for major innovations in the treatment of Wilms tumor.

The initial goals of the group were to:

1. Improve the survival of children with Wilms tumor and other renal tumors
2. Study the long-term outcome of children treated successfully by identifying the adverse effects of treatment
3. Study epidemiology and biology of Wilms tumor
4. Make information regarding successful treatment strategies for Wilms tumor available to physicians around the world

SURGERY

Standard treatment for Wilms' tumor is surgery and chemotherapy

Surgery mainly nephrectomy.the various types of nephrectomy include :

- Simple nephrectomy –the entire kidney is removed .
- Partial nephrectomy- involves the removal of the tumour and part of the kidney tissue surrounding it.
- Radical nephrectomy-most commonly used.in this, the kidney and surrounding tissues,including the ureter and adrenal gland are removed .neighboring lymph nodes may also be removed .

The first goal is to remove the tumor from the involved kidney or major site, even if the cancer has spread (metastasized) to other parts of the body. Sometimes, the tumor can be too large to remove immediately and may have spread into nearby blood vessels, other vital structures, or may be found in both kidneys. In these patients, physicians sometimes use chemotherapy to shrink the tumor before removing it later in the course of therapy. [19-22]

Surgical approaches for various Wilms tumor conditions

	Radical nephrectomy if opposite kidney is normal and functional.
Unilateral disease	Radical nephrectomy: ▪ Mobilize and reflect colon medially to preserve blood supply ▪ Remove perirenal fascia and adrenal gland without opening Gerota's fascia ▪ Excise tumor-bearing kidney with capsule intact. For tumors deemed inoperable at surgical exploration, open biopsy is obtained.
Bilateral disease	Incisional biopsy of both kidneys for surgical staging ▪ each kidney is staged separately ▪ No nephrectomy at time of initial surgery unless very advanced unilateral disease. Definitive surgery after chemotherapy: • unilateral radical nephrectomy and partial nephrectomy on contralateral side • bilateral partial nephrectomy • unilateral nephrectomy only if response is complete on other side
Unresectable disease	-Biopsy - Margins of possible resection, residual tumor or suspicious nodes are outlined with titanium clips - Deferred nephrectomy until after chemotherapy
Renal vein involvement	- En bloc excision of the tumor and thrombus - Does not increase morbidity
Adjacent organ involvement	- Initial surgery limited to biopsy - Chemotherapy for tumor shrinkage - Second-look surgery: ▪ wedge resection of infiltrated organ ▪ downgrades tumor from stage III to II, therefore, reducing subsequent therapy
Metastases	Persistent lung metastasis - tumor on pleural surface of lung ▪ simple wedge resection - scattered superficial lesions ▪ multiple small resections - Deep-seated lesions ▪ anatomical dissection of lobe or segment

Risk factors for surgical complications

- Intravascular extension of tumor into inferior vena cava
- Nephrectomy performed through flank
- Advanced local stage disease
- Resection of other organs
- Tumor diameter >10cm

Complications of wilms tumour surgery

- Intestinal obstruction
- Intraoperative hemorrhage
- Other visceral organ injury
- Vascular complication
- Wound infection, hernia.

CHEMOTHERAPY

Chemotherapy is systemic therapy which involves the use of certain anti-cancer drugs. Chemotherapy is given to all children with Wilms tumor. It may be given:

- To shrink a tumor too large to remove surgically
- After surgery to destroy any tumor cells that might be circulating in the body (called adjuvant therapy)
- To treat cancer that has spread to organs beyond the kidney

Chemotherapy medications are injected into a vein in different combinations and dosages at different times, depending on the type and stage of Wilms tumor. Chemotherapy is ordered by the pediatric oncologist and is usually given by a nurse.

Chemotherapy Treatment Regimes

Tumor and Stage	Tumor and Stage
Focal anaplastic WT • Stage I-III Diffuse anaplastic WT • Stage I	Regimen DD-4A (vincristine/dactinomycin/doxorubicin x 25 weeks; Flank RT)
Focal anaplastic WT • Stage IV Diffuse anaplastic WT • Stage II-III Diffuse anaplastic WT • Stage IV • without measurable disease	Regimen UH1 (cyclophosphamide/carboplatin/etoposide; vincristine/doxorubicin/cyclophosphamide; x 30 weeks; RT)

RADIATION THERAPY

Radiation therapy uses high-energy beams to kill cancer cells. External beam radiation therapy focuses energy onto the cancer using a radiation source outside the body. This type of radiation therapy is often used along with surgery in more advanced cases of Wilms tumor (stages III, IV and V) that have spread beyond the kidney or are not able to be completely removed at surgery. It is used in Stage I and II disease only if there is evidence of unfavorable histology.

The indications for RT were

- Stage III : FH Wilms tumor
- Relapsed Stage I FH Wilms tumor (all relapsed patients with pulmonary or intra abdominal tumor bed recurrence).

Site	Indications & RT Dose
Flank RT	Stage III - 1080 cGy in 6# Stage III local tumor spillage 1080 cGy in 6# Flank or peritoneal biopsy, open biopsy, flank surgical spillage during surgery Recurrent Wilms tumor - 1080 cGy in 6#
Flank RT	Stage III - 1080 cGy in 6# Stage III local tumor spillage 1080 cGy in 6# Flank or peritoneal biopsy, open biopsy, flank surgical spillage during surgery Recurrent Wilms tumor - 1080 cGy in 6#

A boost of 1080 cGy was given to areas of residual tumor after surgery.

TREATMENT OF RECURRENT WILMS' TUMOR

an unresolved question regarding the treatment of recurrent Wilms' tumor is the benefit of high-dose therapy with autologous stem cell rescue. Several groups have reported promising salvage rates resulting from the use of either single or tandem stem cell transplants . It is unclear, however, whether outcomes of high-dose therapy are superior to those obtained with modern, multiagent chemotherapy regimens

A great deal of progress has been made in treating Wilms tumor over the last 25 years. Nine out of ten children are successfully treated with surgery, chemotherapy and sometimes radiation therapy.

Many improvements in treatments have resulted from the work of the Children's Oncology Group (formerly the National Wilms Tumor Study Group).

STANDARD RISK

Patients given dactinomycin, doxorubicin and vincristine and were NOT given radiation therapy (RT)

- **Stage I:** FH, Wilms tumors with loss of heterozygosity for 1p and 16q, AND either

age more than 2 years or tumor weight more than 550 g.

- **Stage II**: FH, Wilms tumor with any weight of tumor or any patient age

Patientsgiven dactinomycin, doxorubicin a nd vincristine PLUS RT

- If they have no loss of heterozygosity for 1p and 16q and have Stage III, FH Wilms tumor.
- All Stage I and II patients with spill (including local needle biopsy) were reclassified as stage III based on NWTS 5 data showing inferior relapse free survival (RFS) for Stage II patients (4yr RFS 70% with spill versus 84% without spill).

PREVENTION

Currently ,there is no known way to prevent wilm's tumour.The risk of many adult cancers can be reduced with certain lifestyle changes (such as staying at a healthy weight or quitting smoking), but at this time there are no known ways to prevent most cancers in children.

The only known risk factors for Wilms tumors (age, race, gender, and certain inherited conditions) cannot be changed. There are no known lifestyle-related or environmental causes of Wilms tumors, so at this time there is no way to protect against most of these cancers.

In some very rare cases, such as in children with Denys-Drash syndrome who are almost certain to develop Wilms tumors, doctors may recommend removing the kidneys at a very young age (with a donor kidney transplant later on) to prevent tumors from developing.

Wilms' tumor can't be prevented. If the child has signs and symptoms that increase the risk of Wilms' tumor, the child's doctor may recommend periodic kidney ultrasounds to look for kidney abnormalities. While this screening can't prevent Wilms' tumor, it may help detect the disease at an early stage, when treatment is most likely to be successful.

PROGNOSIS

Prognosis for Wilms tumor patients is good with at least a 90% cure rate for localized disease and over 70% for metastatic disease with conventional treatment modalities. The overall survival rate of patients is rising steadily mostly due to NWTS collaborative trials.

Factors predictive of outcome:

- Stage
- Histology
- Gene expression profile
- Tumor size
- Age

Relapse free survival of children diagnosed at a younger age in both unilateral and bilateral disease is better than older children. There is a weak correlation between increasing tumor size and adverse prognosis.

Poor prognostic factors:

● anaplasia in stage ii-iv tumors; diffuse anaplasia is worse than focal anaplasia, but even small foci are associated with poor prognosis due to chemotherapy resistance

● high stage (most epithelial-predominant tumors are stage i; most blastema-predominant tumors are stage iii/iv)

● age > 2 years

● large size

PATIENT EDUCATION

As wilms tumour mostly affects the children,most of the care is provided by the parents.

Parental education

Caring for Your Child

As much as parents long to have their child out of the hospital, they often feel unsure of whether they can provide appropriate care after their child comes home. The doctors, nurses, and home health services should provide all the information and support needed to help a parent care for a child between hospital visits.

Depending on the treatment regimen (and a child's general health and the doctor's recommendations), appropriate at-home care can vary. Treatment for most children with Wilms tumor is not as intensive as treatment for other cancers (except for more advanced stages) so most kids won't have tremendous restrictions on them.

Most kids undergoing treatment for Wilms don't have special nutritional requirements or need medication for low blood cell counts, as most other cancer patients do. However, parents must watch for signs of distress, like fever, nausea, vomiting, or diarrhea. A child with a high fever should consult a doctor right away.

Once a child is finished with therapy, the care team will provide a schedule of follow-up tests. Chest X-rays or CT scans may be taken every several months. Stage and histology of the cancer will determine the ultrasound schedule. Blood work and a physical exam may be required to check for adverse effects of the treatment.

For kids who relapse (the cancer returns), prognosis and treatment depend on their prior therapy, the cancer's histology, and how long it's been since the last treatment. The longer it's been, the better. There are few late recurrences of Wilms tumor, so remaining cancer-free for at least 2 years after treatment is generally a very good sign.

Prenatal Testing

If the WT1 germline mutation in the parent has been identified, prenatal diagnosis for pregnancies at 50% risk is possible by analysis of DNA extracted from fetal cells obtained by amniocentesis (usually performed at ~15-18 weeks' gestation) or chorionic villus sampling (usually performed at ~10-12 weeks' gestation). The risk of Wilms tumor developing in a child with a known WT1germline mutation depends on the penetrance of the specific mutation.

Note: Gestational age is expressed as menstrual weeks calculated either from the first day of the last normal menstrual period or by ultrasound measurements.

Pre implantation genetic diagnosis may be an option for some families in which the disease-causing germ line mutation has been identified. []

CASE STUDY

A 7 yrs old female child was admitted in a tertiary care hospital with the chief complaints of palpable abdominal mass (which was detected by her mother), severe abdominal pain, decreased activity, nausea,vomiting, diarrhoea and fever .

Coming to the history, Patient's Birth history revealed an uncomplicated full-term vaginal delivery. This child had no allergies, daily medications, or herbal supplements. Her development was normal, immunizations were up-to-date, and her health had been good. Family-social history consisted of a working mother and father, living in a house in the suburbs outside of a major city. Neither were smokers. There was no family history of neoplasms, gastrointestinal problems, seizure, congenital heart problems, hemoglobinemias or other major illnesses.

On physical exam the patient appeared worrisomely "sick". She was fatigued and completely indifferent to this examiner as well as the uncomfortable aspects of the exam, unlike most children her age. Her temperature was 101.6 degrees Fahrenheit axillary, pulse regular 122 beats per minute, and respirations 36 breaths per minute. Blood pressure was not obtained.

The chest revealed a gallop without murmur heard over the apex at the left fifth intercostal space, midclavicular line, more than likely due to the high-output state of fever and anemia, depicted later in the laboratory data.

An ultrasound examination of the abdomen revealed an echogenic mass in the left kidney measuring 9 × 4 cm. the patient's tumour histology was luckily favorable. A diagnosis of WILMS TUMOUR STAGE –III was made and nephrectomy was performed.

Then the patient was treated with 12 cycles of chemotherapy with the drugs vincristine (1 mg)and cyclophosphamide (200 mg) ,both the drugs are taken parenterally(intravenous route) followed by the 30 cycles of radiation therapy .After the treatment ,the child condition is normalized ,and she was able to do the things on her own but in order to screen the patient the physician recommended the US abdomen to be performed every month .

DISCUSSION

Wilms' tumor was the first solid malignancy most commonly seen in the childhood .Multimodality treatment has resulted in a significant improvement in outcome from approximately 30% in the 1930s to more than 85% in the modern era. Although the National Wilms' Tumor Study Group and the International Society of Pediatric Oncology differ philosophically regarding the merits of preoperative chemotherapy, outcomes of patients treated with either up-front nephrectomy or preoperative chemotherapy have been excellent. The goal of current clinical trials is to reduce therapy for children with low-risk tumors, thereby avoiding acute and long-term toxicities. At the same time, current clinical trials seek to augment therapy for patients with high-risk Wilms' tumor, including those with bilateral, anaplastic, and recurrent favorable histology tumors. Because of the relative rarity of this tumor, all patients with Wilms tumor should be considered for entry into a clinical trial.

CONCLUSION

Wilms tumour is almost curable in majority of the cases. Therefore, it behooves the clinician to perform abdominal exams on routine and episodic visits. Multidisciplinary approach with a good team work of surgeon, pediatric urologist, pediatric oncologist, pathologist and radiotherapist and an adherence to standard protocols is essential to get cure.

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