International Journal of Research and Review
Vol.8; Issue: 3; March 2021
Website: www.ijrrjournal.com
Case Report *E-ISSN: 2349-9788; P-ISSN: 2454-2237*

A Case Report on Nephroblastoma (Wilms Tumour) with Pulmonary Lobe Metastasis

Asif A Latheef[1], Dr. Krishna Kumar Sharma[2]

[1]Department Pharmacy Practice, National Institute of Pharmaceutical Education and Research (NIPER)
[2]Export Promotion Industrial Park (EPIP), Industrial Area, Hajipur - 844 102, Dist: Vaishali, Bihar, India

Corresponding Author: Asif A Latheef

ABSTRACT

A malignant mixed tumour that includes metanephric blastoma, stromal and epithelial derivatives is a Wilms tumour. There are chromosome 11 WT1 gene mutations and nephro-blastematosis (renal blastema persistence in kidney tissue). [1] The tumour is an epithelial part of the tumour (abortive tubules and glomeruli) surrounded by metanephric blastema and immature stroma of the tumour spindle cell. Possible metastases in bilateral upper, middle & right lobe nodular opacities. [2] The case was diagnosed as Left Kidney Nephroblastoma with pulmonary lobe metastasis, based on clinical tests and biopsy techniques. The possible 4th stage of the Wilms tumour.

Key words: Wilms tumour, Nephroblastoma, Nephrectomy, Metastasis

INTRODUCTION

Cancer is a broad group of diseases that, when abnormal cells develop uncontrollably, may start in almost any organ or tissue of the body, go beyond their natural limits to invade adjoining parts of the body and/or spread to other organs. The most prominent childhood abdominal malignancy is the Wilms tumour, or nephroblastoma. Adenosarcoma in the kidney area is the Wilms tumour. The tumour emerges from the remaining bits of embryonic tissue after birth. In the kidney region, this tissue can spark rapid cancerous development.[2]

Stages *[2]*

The Staging of Wilms Tumour has been described by the Children's Oncology Community as:

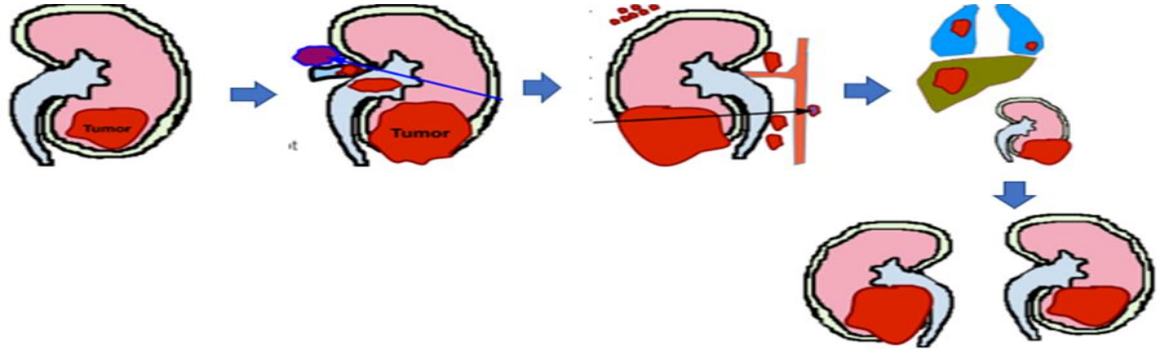
Stage 1. The tumour is confined to the kidney and is fully resected; the renal capsule is intact; the tumour has not been ruptured or biopsied until removal; there is no involvement of the renal sinus vessels and there is no sign of tumour at or outside the resection margins.

Stage 2. The tumour is completely resected; there is no evidence of a tumour at or beyond the resection margins; and the tumour continues beyond the kidney (penetration of renal capsule, involvement of renal sinus).

Stage 3. Following surgery, a residual, non-hematogenous tumour is present and is limited to the abdomen; positive lymph nodes are noted in the abdomen or pelvis; Observed penetration through the peritoneal surface; Presence of peritoneal implants; Postoperatively, gross or microscopic tumour persists, including positive margins of resection; tumour spillage, including biopsy, is noted.

Stage 4. Hematogenous metastases outside the abdomen or pelvis (e.g., lung, liver, bone, brain) or lymph node metastases are noted.

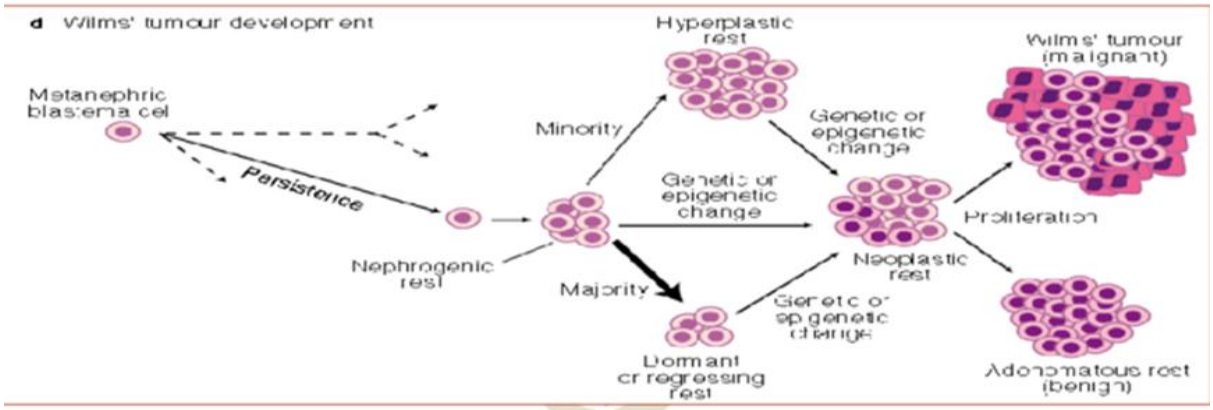
Stage 5. At diagnosis, bilateral renal involvement of tumour is present.



(fig.2-Different stages of Wilms tumour).
(ref;-Sushmita Ghoshal, Management of Wilms tumour, Slide share, Santam Chakraborty, Published on April 07,2007. Multimodality management of Wilms Tumor, Radiation Oncologist at Malabar Cancer Centre, Published in: Health & Medicine, Technology).

Pathophysiology

A malignant mixed tumour containing metanephric blastoma, stromal, and epithelial derivatives is the Wilms tumour. [7] WT1 gene mutations on chromosome 11 and nephronic blastematosis (persistence of renal blastema in kidney tissue) have been reported. The tumour consists of the tumour epithelial part surrounded by metanephric blastema and tumour immature spindle cell stroma (abortive tubules and glomeruli). Differentiated (muscle, cartilage, bone, fat tissue, fibrous tissue) or anaplastic elements may be found in the stroma. The usual kidney parenchyma is squeezed by the tumour. [5]

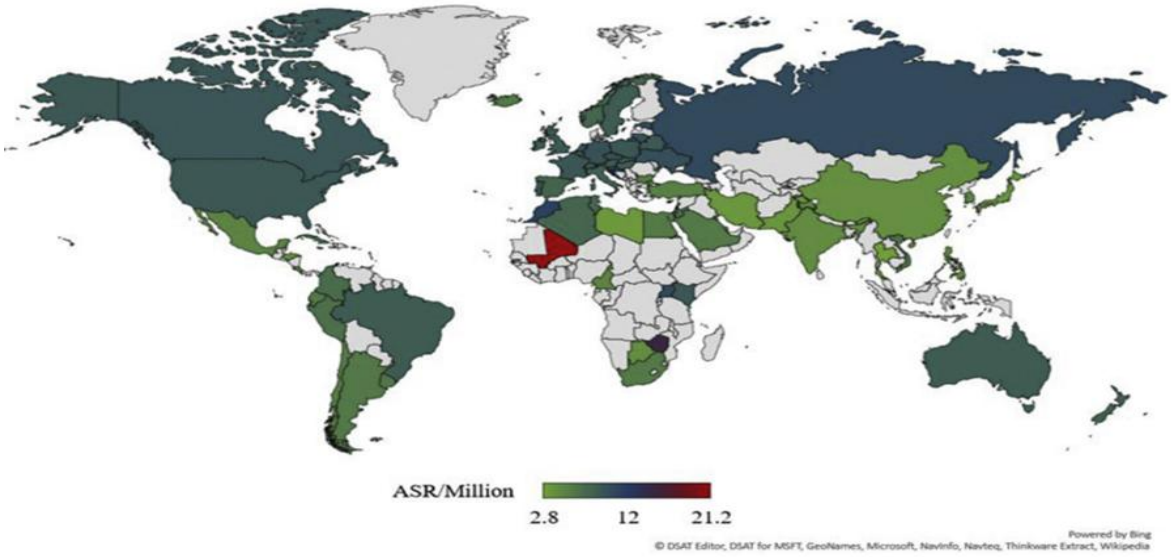


(fig.3-pathophysiology of WT)
((ref;-Sushmita Ghoshal, Management of Wilms tumour, Slide share, Santam Chakraborty, Published on April 07,2007. Multimodality management of Wilms Tumor, Radiation Oncologist at Malabar Cancer Centre, Published in: Health & Medicine, Technology).

Epidemiology and Incidences

In the United States and around the world, the incidence of the Wilms tumour is as follows:

Around 10 children and adolescents per 1 million before the age of 15 years are affected by the Wilms tumour. [7] The Therefore, it accounts for 6-7 percent of all North American childhood cancers. As a result, on this continent, about 450-500 new cases are diagnosed per year. For unilateral impairment, the male to female ratio is:- 0.92:1.00. [7] The Wilms tumour tends to be more prevalent in Africa, and less prevalent in East Asia. In blacks, Wilms' tumour is comparatively more prevalent than in whites and is uncommon in East Asians. Estimates indicate that there are 6-9 cases per million person-years among whites, 3-4 cases per million person-years among East Asians, and more than 10 cases per million person-years among black people. [4] The male to female ratio was 0.92 among patients with unilateral Wilms tumour enrolled in all NWTSG protocols: [8].



(fig.4-Epidemiology and incidence of Wilms tumour).
(ref;- Kaatsch P, Grabow D, Spix C. „German Childhood Cancer Registry-Jahresbericht/Annual Report 2016 (1980-2015)

Causes

The Wilms tumour is believed to be caused by gene changes responsible for the WT1 gene's normal genitourinary growth. As a direct result of the research of children with Wilms tumour who also had aniridia, genitourinary abnormalities, and mental retardation, WT1, the first Wilms tumour suppressor gene at chromosomal band 11p13, was established (WAGR syndrome). [2] A second gene predisposing individuals to Wilms tumour was identified (but not yet cloned) as telomeric WT1 at 11p15; this locus was suggested on the basis of studies in patients with both Wilms tumour and Beckwith-Wiedemann syndrome (BWS), another chromosomal band 11p15 congenital Wilms tumour predisposal syndrome. [3]

Symptoms

- Classically appears as a silent abdominal mass during childhood (60-70%)
- Other symptoms:

– Pain Abdomen (30-40%)
– Flank pain and rapid enlargement of the mass (2° to bleeding in the tumor)
– Hematuria (25%)
– Fever (20%)

SUBJECTIVE FINDINGS

Patient Details

A 5 year old F/patient was came to the hospital with heterogenous lobulated mass arising from left kidney (CECT of whole abdomen).

Presenting Complications

- Bulging of abdomen at left side.
- Fever.
- Pain abdomen.
- Loss of appetite and fatigue.
- Pallor and lymphadenopathy present.

Examinations Suggested

- General examinations.
- Haematology examinations.
- Biochemical examinations.
- CT-scan of thorax + whole abdomen NCCT&CECT (non-ionic),
- FNAC Biopsy and Biopsy large.

General examinations *shows;*

- Body weight :-17 kg.
- BP :-100/60 mmHg.
- Pulse :-150/min.
- Pallor present.
- Lymphadenopathy present.

HAEMATOLOGICAL REPORTS (*table 1)*												
SL NO	INVESTIGATIONS	NORMAL	16/10/19	6/11/19	20/11/19	11/12/19	25/12/19	15/01/2020	26/01/20	13/02/20	24/02/20	9/03/20
1	WBC (10^3/mm^3)	4-11	10.2	15.5	5.5	4	3.3	16.4	5.5	2.8	1.1	3.9
2	RBC (10^6/mm^3)	3.5-5.5	3.16	3.44	3.65	3.58	3.02	3.4	3.34	3.29	3.04	3.64
3	HGB(g/dl)	14-18	8.4	9.3	10.1	10.3	9.2	9.6	9.5	9	8.3	10
4	HCT(%)	40-55	27.7	30.5	32.2	31.4	26.5	29.7	29.3	27.7	25.2	31
5	MCV (FI)	80-100	87.66	88.6	88.2	87.71	87.75	87.35	84.72	84.19	82.89	85.1
6	MCH(pg/ml)	27-37	26.5	27	27.6	28.72	30.46	28.24	28.44	27.36	27.30	27.7
7	n(MCHC) (g/dl)	32-36	30.3	30.4	31.3	32.8	34.72	32.32	32.42	32.49	32.94	32.5
8	PLT (10^3/mm^3)	150-140	220	298	130	88	110	129	97	62	102	216
9	MPV(fL)	7.4-10.4	8.6	9.1	11	9.9	10.4	10.1	8.3	7.9	8.3	10.1
10	PWD(Fl)	15-17	15.8	16.5	24.3	21.8	24.5	21.5	13.8	14.5	14.5	22.3
11	PCT(%)	0.1-0.28	0.23	0.27	0.09	0.057	0.072	0.13	0.078	0.05	0.056	0.21
12	Neutrophils	40-70%	66.4	59.6	72.8	51.1	50.1	79.4	56.9	75.5	31.4	73
13	Lymphocytes	20-40%	26.5	21.6	64	17.6	32.1	10.9	24.1	16.5	42.6	14.7
14	Eosinophil	1-4%	0.9	2.4	12.8	12.8	4.1	2.5	5.5	5.6	7.6	4
15	Monocytes	2-8%	5.7	7.6	7.5	17.4	13.1	6.4	12.5	1.9	15.4	2
16	Basophiles	0-1%	0.5	0.8	0.57	1.1	0.6	0.8	1	0.5	3	1.4
17	ALY	0-2.5%	0.8	1.2	1.4	3.3	1.2	0.7	1.5	0.4	2.5	2
18	LIC	0-3	0.8	0.9	1.5	1.7	1.4	2.9	0.8	0.5	3.2	6.9

Biochemical investigations;-

LIVER FUNCTION TESTES *(table 2)*		
TEST	**REFERENCE**	**OBSERVED**
Bilirubin(total)(mg%)	0.2-1.2	**0.32**
Bilirubin (direct)(mg%)	0-0.4	**0.11**
Bilirubin(indirect)(mg%)	0.1-0.6	**0.21**
SGOT(AST)(U/L)	10-35	**37**
SGPT(ALT)(U/L)	9-43	**8**
ALP(U/L)	185 -499	**123**
Protein(g%)	6.5-8.5	**5.6**
Albumin(g%)	3.5-5.3	**2.8**
Globulin(g%)	1.8-3.6	**2.8**
A/G Ratio	1-2.1	**1**

RENAL FUNCTION TESTES *(table 3)*		
TEST	**REFERENCE**	**OBSERVED**
Blood urea(mg%)	13-50	**23.54**
BUN(mg%)	6-21	**11**
Creatinine(mg%)	0.6-1.3	**0.44**

ELECTROLYTES *(table 4)*		
TEST	**REFERENCE**	**OBSERVED**
Sodium (m eq/L)	130-145	**122**
Potassium (m eq/L)	3.5-5.5	**4.9**
Chloride (m eq/L)	97-111	**105**

Ct Scan Of Thorax +Whole Abdomen NCCT & CECT (Non-Ionic);-

Nodular opacities in bilateral upper, middle & right lobe likely metastasis.

Expansile heterogeneously enhancing soft tissue density lesion arises from left kidney replacing it with areas of necrosis and calcification with extra renal spread, partially enhancing aorta and is displacing it towards the right side.

Lesion is abutting tale of pancreas and bowel loop anteriorly laterally abutting intero-medial aspects of spleen and lateral abdominal wall. posteriorly infiltrating left poas muscle extending in left para vertebral region at the same level. Left adrenal gland not visualised separately.

FNAC Biopsy; -Adenocarcinoma of kidney with lymph node involvement. (Specimen of
a) left radical nephrectomy
b) inter aortocaval node
c) omental nodule)

Diagnosis

Based on the clinical examinations and the biopsy techniques, the case was diagnosed as Left kidney nephroblastoma with pulmonary lobe metastasis. Likely 4th stage of Wilms tumour.

Patients History

A 5 Year old female patient were came to the hospital with heterogenous lobulated mass arising from left kidney (CECT of whole abdomen).

Nodular opacities in bilateral upper, right middle lobe likely metastasis

Expansile heterogeneously enhancing soft tissues arises from kidney and replaces with extrarenal spread (partially encasing aorta and by right side).

CT scan of thorax NCCT & CECT (non ionic)

Lesions were seen: -Anterior (abutting tale of pancreas & bowel loop)

Laterally (inferomedial aspect of spleen & abdominal wall)

Posteriorly (infiltrating:left poas muscle &extending left para vertebral region)

FNAC done for further evaluation wilms tumor of left kidney.

Chemotherapy started from (first cycle)

After 6 cycles of therapy again taken CT-scan of thorax + whole abdomen NCCT&CECT(non-ionic) shows: Tumor size (14*8*7 cm)

No focal lesion in lungs, bowel loop etc. spleen & pancreas became normal. Vitals were normal up to that.

Radical nephrectomy were carried out at 8th cycle of chemotherapy. shifted to recovery room. A Specimen taken by operation is send for biopsy (large).

Received from recovery room.

Stich removed and Radiotherapy started.

Treatment Plan

Chemotherapy started with,

Rx,

- ✓ VINCRISTINE-1.1mg IV injection
- ✓ ACTINIMYCIN-0.7 mg IV injection (3 cycle)
- ✓ DOXORUBICIN-35mg IV injection (1 cycle)

Vincristine used up to 8 cycles after that RADICAL NEPHRECTOMY of left kidney done.

Collected samples were undergoes biopsy.

After 13 days of surgery initiate radiotherapy.

Reduction of dose of VCR after checking chemotherapy induced changes (10th cycle (0.72 mg) and 13th cycle (0.34mg).

Radiotherapy

Radio therapy were initiated from 9/01/2020;

Type- Three dimensional conformal radiation therapy. (3DCRT).

machine: linear accelerator.

Position; supine.

Plan; EBRT to thorax+flank-external beam radiation therapy.

Patient immobilization device-thermoplastic mould on pelvis.

Base plate on pelvis-Carbon fiber.

RADIOTHERAPY CHART

(table 6)

No	1	2	3	4	5	6	7	8
Date	9/01/2020	10/01/2020	11/01/2020	13/01/2020	14/01/2020	15/01/2020 2^{nd}dose stopped	16/01/2020	17/01/2020 First dose Stopped.

Summary of Patient

A 5 year old female child was admitted with left nephroblastoma with pulmonary metastasis.

Chemotherapy initiated and surgery done.

The biopsy after surgery; -

(a) radical nephrectomy
(b) interaotocaval node
(c) omental nodule shows

1)Necrosis seen.
2)Capsule is free of tumour. } histopathology
3)Ureter and vessels are free of tumour.
4)Lymph o vascular invasions absent.
5)Lymph nodes:- 6 lymph nodes negative for tumour metastasis (a)
3 lymph nodes negative for tumour metastasis (b)
1 lymph node is negative for tumour metastasis (c)

DISCUSSION

The patient was admitted with fourth stage of Wilms tumour with pulmonary metastasis. She was undergoes chemotherapy after that surgery done and radiotherapy also was done. At the end the condition of patient were better the lobes were free from metastasis. It indicates the positivity in treatment. For avoiding the chemotherapy as well as the radiotherapy induced adverse events the interventions for efficacy and safety became implemented.

CONCLUSION

The study conducted and to check the new treatment strategies for childhood tumour and its complication. In this particularly study the patient were at fourth stage of Wilms tumour with pulmonary lobe metastasis. By the normal chemotherapy regimen the patients was free from the metastasis and after the surgery the mass removed and radiotherapy initiated at the end positive results was observed.

REFERENCE

1. Megan E Cunningham, Theodore D Klug, Jed G Nuchtern, Murali M Chintagumpala, Rajkumar Venkatramani, Joseph Lubega, Bindi J Naik-Mathuria Global Disparities in Wilms Tumor. 2020 Mar; 247:34-51.
2. Maria Yiallouros, Editor: Maria Yiallouros, Release: Wilms Tumour (Nephroblastoma) Brief information Prof. Dr. med Wilms Tumour. Author created 12/10/2009.
3. S1-Leitlinie. Graf N Nephroblastoma (Wilms-Tumor) AWMFonline2016 025/004AWMF025-004l.
4. Graf N, Rübe C, Gessler M „Nierentumoren, in: Gadner H, Gaedicke G, Niemeyer CH, Ritter J (Hrsg.): Pädiatrische

Hämatologie und Onkologie, Springer-Verlag 2006: 847-865,3540037020.
5. Graf N, Semler O, Reinhard H „Prognosis of Wilm's tumor in the course of the SIOP trials and studies“, Urologe A 2004;43:421-428
6. Kaatsch P, Grabow D, Spix C. „German Childhood Cancer Registry-Jahresbericht/Annual Report 2016 (1980-2015)
7. Claudia Spix,2 Peter Kaatsch,2 and Fritz Lampert3 ncidence, Survival, and Treatment of Localized and Metastatic Neuroblastoma in Germany 1979–2015. Published online 2017 Aug 7. doi: 10.1007/s40272-017-0251-3 PMCID: PMC5680382.
8. M. Gleason, Armando J. Lorenzo, Paul R. Bowlin, and Martin A. Koyle. Innovations in the management of Wilms' tumorM 2014 Aug; 6(4): 165–176.doi: 10.1177/1756287214528023.
9. Sushmita Ghoshal, Management of Wilms tumour, Slide share, Santam Chakraborty, Published on April 07,2007. Multimodality management of Wilms Tumor, Radiation Oncologist at Malabar Cancer Centre, Published in: Health & Medicine, Technology).

How to cite this article: Latheef AA, Sharma KK. A case report on nephroblastoma (wilms tumour) with pulmonary lobe metastasis. *International Journal of Research and Review*. 2021; 8(3): 212-218.

******