

Management of Wilms' Tumor

Essay submitted for partial fulfilment
of M.S. degree in Radiation,
Oncology and Nuclear Medicine

by
Ihab Mostafa Mohamed
M.B., B.Ch.

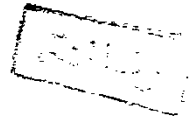


Supervisors

Dr. Atef Yousef Ryad
Assis. Professor of Radiation
Oncology and Nuclear Medicine
Faculty of Medicine
Ain Shams University

Dr. Soheir Helmy Mahmoud
Assis. Professor of Radiation
Oncology and Nuclear Medicine
Faculty of Medicine
Ain Shams University

Dr. Soheir Said Ismail
Lecturer of Radiation,
Oncology and Nuclear Medicine.
Faculty of Medicine
Ain Shams University



Ain Shams University

Cairo

1991

Acknowledgement

First and foremost thanks are to Allah, The most Beneficient and Merciful.

I am really indebted to Dr. Atef Yousef, assisstant professor of Radiotherapy and Nuclear Medicine, Ain Shams University, for accepting supervision, unlimited support and kind help throughout this work.

I would like also to express my cordial thanks and special gratitude to Dr. Soheir Helmy, Assisstant Professor of Radiotherapy and Nuclear Medicine, Ain Shams University, for her patience and judicious guidance; her criticism was always stimulating and encouraging.

And to Dr. Soheir Ismail, Lecturer of Radiotherapy and Nuclear Medicine, Ain Shams University, my deep appreciation is extended, for her meticulous work and close supervision.

Last but not least, I acknowledge all those who helped me and shared in achieving this work.

Ihab Mostafa



This Work Is Dedicated To

My Parents

&

My Brother

Contents

	Page
- List of abbreviations	1
- Introduction	2
- Anatomy	4
- Embryology	7
- Epidemiology	13
- Pathology	22
- Clinical picture	37
- Investigations	40
- Staging	61
- Differential Diagnosis	63
- Treatment	68
Surgery	68
Chemotherapy	73
Radiotherapy	85
- Integration of Treatment	100
- Bilateral Wilms' Tumor	118
- Prognosis	131
- Summary	137
- References	141
- Arabic summary	

List of Abbreviations

List of Abbreviations

ADR	Adriamycin.
AMD	Actinomycine D.
CCS	Clear cell sarcoma.
CPM	Cyclophosphamide
CT	Computerized Tomography.
CTH	Chemotherapy.
CXR	Chest X-Ray.
FH	Favourable Histology.
IVP	Intravenous Pyelography.
MRI	Magnetic Resonance Imaging.
NWTS	National Wilms'Tumor study
RTH	Radiotherapy.
SIOP	International Society of Pediatric Oncology.
U/S	Ultrasonography.
UH (UFH)	Unfavourable Histology.
VCR	Vincristine.

Introduction

Introduction

Wilms' tumor is a highly malignant tumor of mixed histology. It is the most common internal malignant tumor of childhood and the second most common intra abdominal non lymphomatous malignancy in childhood.

Wilms' tumor serves as a model for management of other pediatric tumors in classification, staging, approach to treatment, prognosis, doubling time, results, complication, and late sequelae.

Success story of Wilms' tumor is the ability of chemotherapy to control both overt and occult metastases. This achievement initiates confidence in combination modality approach, also large tumor, regional lymph nodes were ablated by combination of surgery, irradiation in many different dose prescriptions (Putnam et al., 1983). These together will increase accuracy of early diagnosis and proper evaluation of the patient.

At the begining of 20th Century, Wilms' tumor had an operative mortality rate of 23% (D'Angio et al., 1976), survival rate with surgery alone of 8%. Then during the 1930s survival rate rose to 40% because of improvement in surgical and

anaesthetic management of the patient (Ladd 1938). In the 1950s, addition of radiotherapy to tumor-bed increased survival rate to 50% (Collins 1958). By the 1960s, combination of surgery, radiation and chemotherapy especially actinomycin D, vincristine led to increased survival to 81% (D'Angio 1980). By 1969, several National Co-operative study groups combined with individual institutions to form National Wilms' Tumor Study (NWTs) I, II, III. The goal was to determine what combination of therapies is best for treatment of all types and stages of the disease (D'Angio et al., 1976).

Aim of the work is to revise recent trends in the management of Wilms' tumor and its impact on improving survival.

Anatomy

Anatomy of The Kidney

The kidneys are situated in the posterior portion of the abdomen at a level between the eleventh rib and the third lumbar transverse process. The right kidney is usually situated 2 cm. lower. Anteriorly, the right kidney is in relation to the descending portion of the duodenum, hepatic flexure of the colon and the right lobe of the liver and the left kidney is in relation to the tail of pancreas and the posterior wall of the stomach. The kidney in its fibrous capsule and perinephric fat are enveloped by Gerota's fascia which arise from transversalis fascia (Juan et al., 1977).

Lymphatics: (fig. 1)

Two separate systems of lymphatic channels have been demonstrated in renal parenchyma (Rawsen, 1949). One system begins close to the capsule forms nets about the cortical blood vessels, winds around interlobar and arcuate vessels and leaves the kidney by the hilus. Another system begins beneath the mucosa of the papillae, ascends through the medulla and enlarges to form channels that surround the arcuate blood vessels. At the pedicle the lymphatics divide into three trunks (Rouviere, 1933):

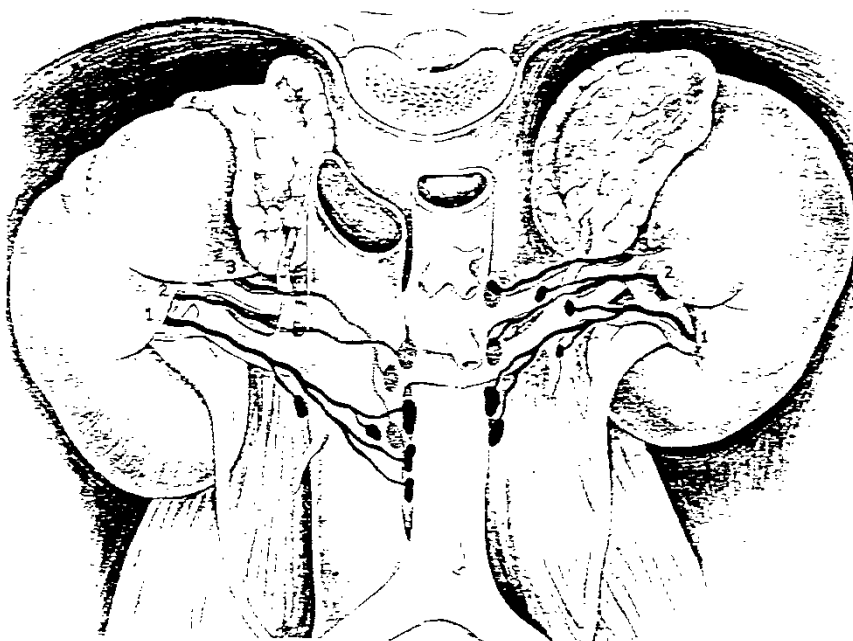


Fig. 1: Anatomic sketch of the lymphatics of the kidneys showing, 1, anterior trunks; 2, middle trunks; 3, posterior trunks (After Rouvière).

- 1- The anterior trunk drains the front half of the kidney and ends in the paraaortic nodes between the renal and inferior mesenteric artery. On the left side they may end also into a node at the junction of suprarenal and renal veins and into a node located at the termination of the spermatic vein in the renal vein. At times they may also terminate in a precaval lymph node.
- 2- The middle trunk terminates in a paraaortic node on the right side and on a node at the junction of the suprarenal and renal veins on the left side.
- 3- The posterior trunk: drains the posterior half of the kidney and terminates in the nodes along the border of the aorta and behind the inferior vena cava on the right side and in paraaortic nodes near the origin of the renal artery on the left side (Juan et al., 1977).

Blood Supply

Renal arteries: are large vessels which run laterally from the sides of aorta opposite the L₁₋₂ intervertebral disc. Because of the slightly asymmetrical positions of the aorta and the two kidneys, the right renal artery is rather longer than the left. Each artery crosses the crus of the diaphragm and as it approaches the hilus

of the kidney it divides into numerous branches. The highest of these branches turns upward to reach the supra renal gland while the lowest turn downward to supply the upper part of the ureter. The other branches pass through the renal hilus into the sinus of the kidney and enter the kidney substance between the minor calyces. The majority of these branches pass in front of the renal pelvis (Smith et al., 1983).

Aberrant renal arteries: these usually arise from the aorta and usually enter the kidney substance below or above the hilus without entering the renal sinus (Smith et al 1983).

Renal veins are formed, on either side, by the convergence of numerous tributaries in the renal sinus. It emerges through the kidney hilus, in front of the renal artery and the renal pelvis. The two veins are wide vessels of very different lengths. The right is only about 1 cm long. The left is about four times as long. After passing to the right in front of the corresponding artery it crosses in front of the aorta. Although the right suprarenal and the right gonadal veins drain directly into the inferior vena cava, on the left side, the corresponding vessels drain from above and below into the left renal vein (Smith et al., 1983).