WILMS TUMOUR

THANA BALAN
INTRODUCTION

• Most common neoplasm in children
• 90% occurs in children< 7 years of age
• Boys and girls are equally effected
• Peak incidence is in children between 3 and 4 years of age
ETIOLOGY

• Nephrogenic rests may be the precursor lesion
• Nephrogenic rests consist of focus of abnormally persistent nephrogenic cells
• Nephrogenic rests is seen in 40% of children of Wilms tumour and approaches 100% in bilateral synchronous tumours
ETIOLOGY

- Wt1 GENEis located in 11p13 chromosome. Defect in this gene is associated with wilms tumour and associated with WAGR and Denys Drash syndrome
ETIOLOGY

- WT2 GENE is located in 11p15 chromosome. The defect of this gene is associated with Wilms tumour and Wiedemann syndrome.
ANOMALIES WITH WILMS TUMOUR

- 15% of children with Wilms tumour has congenital anomalies;
  - aniridia
  - WAGR syndrome
  - hemihypertrophy
  - Beckwith-Wiedemann syndrome
  - Denys Drash syndrome
PATHOLOGY

• Encapsulated tumour
• Necrosis and haemorrhage common
• Tumour rarely grows into renal pelvis
• Invasion into renal veins is seen in 20% of cases
• Microscopically shows triphasic histology containing blastemal, epithelial and stromal cells
PATHOLOGY

• Anaplasia suggests bad prognosis
PRESENTATION

• Abdominal mass which rarely crosses the midline
• Pain due to haemorrhage
• HPT in 70% cases due to increase renin
• Varicocele if tumour propagates into renal veins
INVESTIGATIONS

- u/s
- Ct
- Ivu
- mri
STAGING

• Stage 1- limited to kidney and completely excised
• Stage 2- breach renal capsule and completely excised with local spillage
• Stage 3- node positive, diffuse spillage and positive margins
• Stage 4- metastases
• Stage 5- bilateral tumours
MANAGEMENT

• Multimodality treatment
• Either NWTS or SIOP approach
• NWTS; surgery first then chemo or radiotherapy
• SIOP; chemotherapy first then surgery
MANAGEMENT

• Guidelines in Malaysia;
• - Early resection except stage 5 or unrectable
• - Transperitoneal with complete abdominal exploration
• - Inspect contralateral kidney
• - Partial nephrectomy not recommended
• - Avoid major spillage
• Resectable WILMS tumour
• stage 1 ; sur+ VCR for 10 weeks
• stage 2; sur+ VCR+ACT for 26 weeks
• stage 3+4 ;sur+ VCR+ACT+ADR for15 weeks with radiotherapy
• Indications for preoperative chemotherapy;
• -Stage 5
• -tumour in single kidney
• -extensive IVC thrombus
• - large tumour