# Renal Tumours

## Renal cell carcinoma

# Incidence

**This is primarily a disease of the elderly patient, with typical presentation in the sixth and seventh decades of life**

# Etiology and Environmental Factors of Malignant Renal Tumours

**Established**

Tobacco exposure

Obesity

Hypertension

**Putative**

Lead compounds, aromatic hydrocarbons, Trichloroethylene exposure

Occupational exposure (metal, chemical, rubber, and printing industries)

Asbestos or cadmium exposure

Radiation therapy

# Genetic abnormality in RCC

There is Loss of VHL tumour suppressor gene on chromosome 3 in RCC

Inactivation or mutation of the *VHL* gene leads to dysregulated expression of the Hypoxia Inducible Factors. This leads to a severalfold expression of vascular endothelial growth factor (VEGF), the primary angiogenic growth factor in RCC, contributing to the pronounced neovascularity associated with clear cell RCC

There is an association of RCC with Von Hippel Lindau (VHL) disease & Tuberous sclerosis

# Pathology of RCC

**Most RCCs are round to ovoid and circumscribed by a pseudocapsule of compressed parenchyma and fibrous tissue rather than a true histologic capsule**

**All RCCs are, by definition, adenocarcinomas, derived from renal tubular epithelial cells**

**Clear cell RCC accounts for 70% to 80% of all RCCs**

# Pathology of RCC

Unique feature of RCC is its predilection for involvement of the venous system, which is found in 10% of RCCs.

This is most commonly manifested in the form of a contiguous tumour thrombus that can extend into the inferior vena cava (IVC) as high as the right atrium

# Pathology of RCC

The most common site of distant metastases is the lung.

However, liver, bone (osteolytic), ipsilateral adjacent lymph nodes and adrenal gland, brain, the opposite kidney, and subcutaneous tissue are frequent sites of spread

**TNM classification system for renal cell carcinoma**

**T—Primary tumor**

**T1**

T1a Tumour less than 4.0 cm in greatest dimension, limited to the kidney

T1b Tumour 4.0–7.0 cm in greatest dimension, limited to the kidney

**T2**

T2a Tumour more than 7.0 cm in greatest dimension, limited to the kidney

T2b Tumour >7 cm but <10 cm in greatest dimension, limited to kidney

## TNM classification system for renal cell carcinoma

T3a Tumour invades renal vein or its segmental branches or perirenal fat or renal sinus fat but not beyond Gerota's fascia

T3b Tumour grossly extends into vena cava below the diaphragm

T3c Tumour grossly extends into vena cava above diaphragm or into the wall of the vena cava

T4 Tumour invades beyond Gerota's fascia including contiguous extension into ipsilateral adrenal gland

## TNM classification system for renal cell carcinoma

**N—Regional lymph nodes**
NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in regional lymph nodes

**M—Distant metastases**
MX Distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

# Symptoms & signs of RCC

Classical triad of gross haematuria, flank pain and a palpable mass occurs in only 7–10% of patients and is frequently a manifestation of advanced disease

Patients may present with haematuria, dyspnoea, cough, and bone pain that are typically symptoms secondary to metastases

With the routine use of CT scanning for evaluation of nonspecific findings, asymptomatic renal tumours are increasingly detected incidentally (>50%)

# Symptoms & signs of RCC

**Paraneoplastic Syndromes Associated with Renal Cell Carcinoma**

Elevated erythrocyte sedimentation rate

Hypertension

Anaemia

Cachexia, weight loss

Pyrexia

Hypercalcemia

# Symptoms & signs of RCC

Stauffer syndrome of hepatic dysfunction

Hepatic function abnormalities include elevation of alkaline phosphatase and bilirubin, hypoalbuminemia, prolonged prothrombin time

# Investigations of RCC

Lab Investigations:

CBC, LFT, KFT, Serum Calcium

Radiological Investigations:

Contrast enhanced CT scan is the primary investigation for diagnosis & staging. The typical finding is a mass which enhances on contrast scans

Ultrasound examination is a non invasive, relatively inexpensive technique able to further delineate a renal mass. It is approximately 98% accurate in distinguishing simple cysts from solid lesions. Ultrasonographic criteria for a simple cyst include a well-circumscribed mass without internal echoes, thick septa or nodules which enhance

# Treatment of RCC

Partial or Radical nephrectomy is the treatment for localised disease. For small tumours partial nephrectomy is preferred.

Radical nephrectomy involves removal of the kidney along with gerota's fascia, upper ureter and lymph nodes at the hilum. Regional lymph node dissection is not beneficial as by this time the disease is already metastatic. Thrombus in the renal vein and vena cava is usually free floating and it is possible to remove it with venotomy / cavotomy

Laparoscopic radical or partial nephrectomy has lesser morbidity

# Treatment of RCC

Locally advanced or metastatic disease is treated with cytoreductive nephrectomy and

resection of solitary metastases especially a solitary lung met

Adjuvant therapy:

Vascular growth factor and kinase inhibitors: **Sunitinib is most commonly used**

or Sorafenib

Vascular growth factor antagonists: Bevacizumab

Other drugs tried are Interferon or Interleukin 2

# Prognosis of RCC

Tumour Localised to the kidney has 5 year survival 70% – 90 %

Prognosis worsens with locally advanced disease, lymph node metastases or systemic disease for which 5 year survival is 0 – 20%

# Benign tumours of the kidney

## Renal Angiomyolipoma

# Incidence

Angiomyolipoma accounts for less than 10% of renal tumors

# Incidence

Twenty to 30 percent of angiomyolipoma's are in patients with Tuberous Sclerosis

# Pathology

It consists of thick-walled aneurysmal vessels, smooth muscle, and varying levels of mature adipose tissue

# Presentation

The typical presentation is of a middle-aged woman with a single asymptomatic tumour detected on ultrasound or CT scan

Sporadic angiomyolipoma's appear to have a slow growth rate

The Wunderlich syndrome, or massive retroperitoneal haemorrhage, is the most significant complication of renal angiomyolipoma

# Investigation

Angiomyolipoma is the only benign renal tumour that is confidently diagnosed on cross-sectional imaging. The presence of fat (confirmed on nonenhanced thin-cut CT by a value of −20 Hounsfield Units [HU] or less) within a renal lesion is considered the diagnostic hallmark

# Management

Intervention should be considered for large tumours > 4 cms, particularly if the patient is symptomatic.

Women of childbearing age and patients with limited access to surveillance or to emergency care should also consider a proactive approach in view of the risk of bleeding

# Management

Treatment options for elective management of larger angiomyolipoma’s include selective renal angioembolization and open or minimally invasive partial nephrectomy

# Some Other benign renal tumours

Oncocytoma

Metanephric Adenoma

Leiomyoma