Neuroblastoma

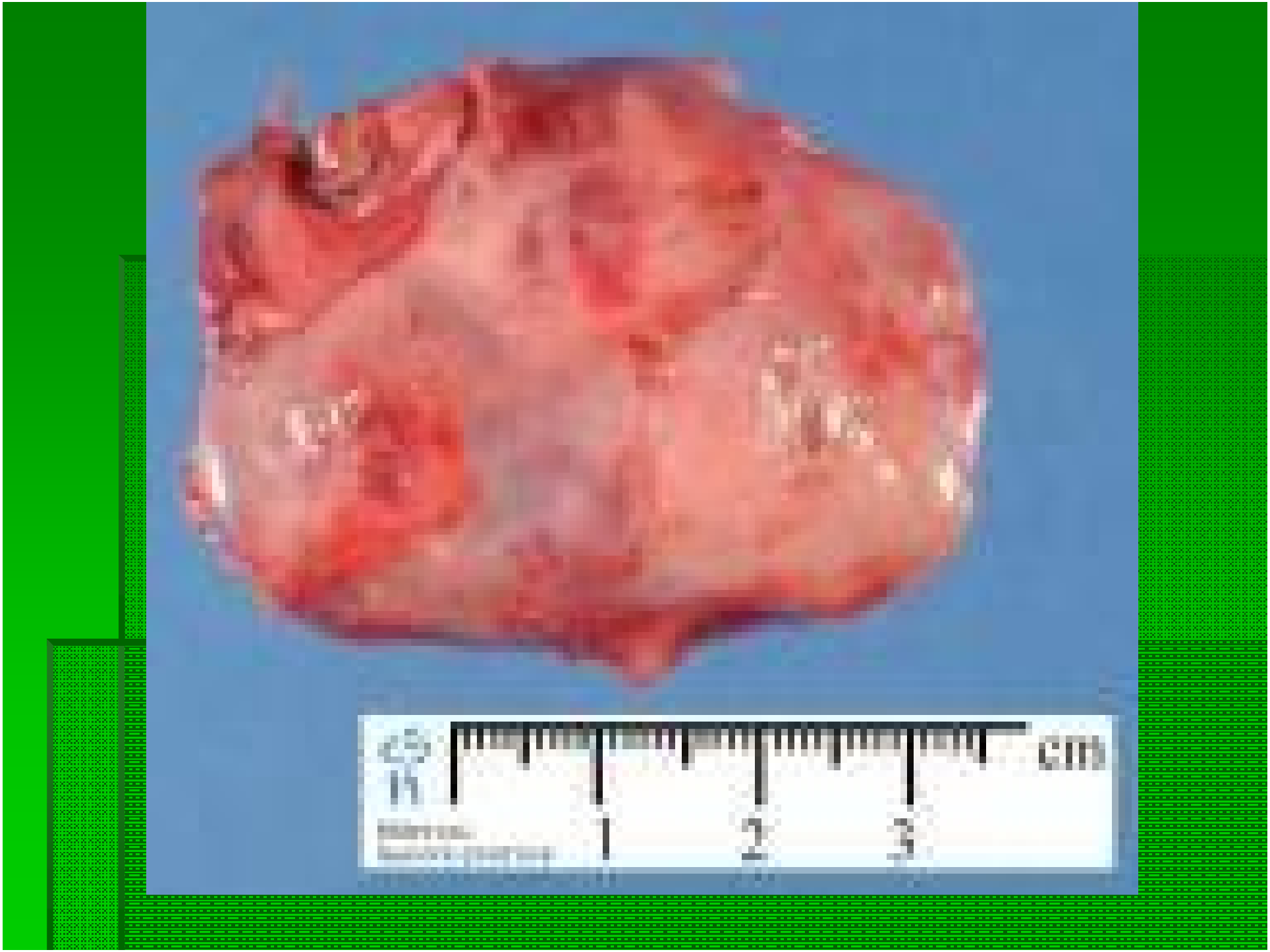
Tumor derived from neural crest cell that form the sympathetic ganglia&adrenal medulla.

Causes

- *unknown.
- *familial neuroblastoma has been reported but is rare .
- * The incidence is 1:100,000 per yr in children under 15 yr.
- *more common in white than black.
- *slightly more common in boy than girl.*medium age at diagnosis is 2Yr.75% of cases<5Yr.

Pathology

- *neuroblastoma is a firm, gray tumor with bleeding,necrosis&calcification.
- *most cell are primitive neural cell without obvious differentiation (small blue round cell)
- *tumor can arise anywhere neural crest cell migrate
- 70% arise in the abdomen.
- 50% of them in the adrenal gland.
- 20% arise in thorax
- 10% arise elsewhere (unknown site



Complication

- Common site of metastases include:
- *liver
- *bone marrow
- *skin
- lymph node.
- *bone particularly the skull .
- _Mass effect at site of primary lesion.

Complication

- -paraneoplastic syndrome:
- Vasoactive intestinal peptide(VIP)
- (Opsoclonus_myoclonus) syndrome.
- (VIP) syndrome; due to (VIP) production; watery diarrhea, abdominal distention &electrolyte imbalance,
- (opsoclonus- myoclonus) unknown causes
- with chaotic eye movement (dancing eye) and myoclonus jerk (dancing feet)
-

Clinical manifestation

- Presenting sign &symptoms of Neuroblastoma depend on the primary site of the tumor and the degree of dissemination.
- *Abdominal mass:
- the mass usually firm,fixed,irregular mass frequently crosses the midline.
- *Abdominal distention with or without tenderness.
- *Mass effect:
- sign of bowel obstruction:(anorexia, vomiting,)
- *Liver metastasis _enlargment&ascitis.
- Cervical/ thoracic mass:
- Neck mass

Clinical manifestation

- *Horner syndrome: ptosis,myosis&anhydrosis..
- *Respiratory distress or stridor.
- *Superior vena cava syndrome with large mediastinal tumors.
- Paraspinal mass:
- *Vertebral body involvement& nerve root compression
- *Bladder&bowel dysfunction&back pain.
- Metastatic disease:
- Bone pain,fever,failure to thrive.
-

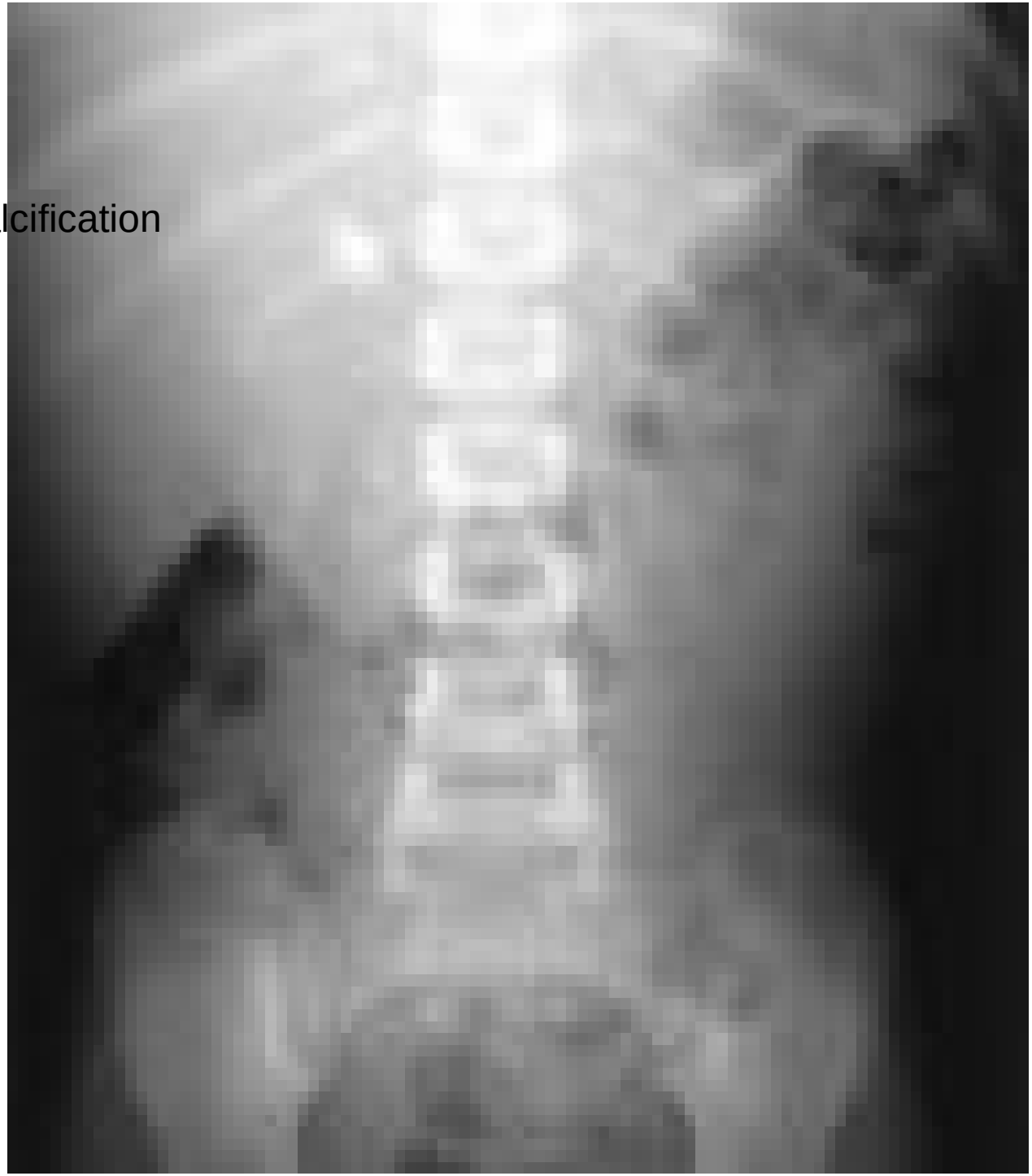
Diagnosis

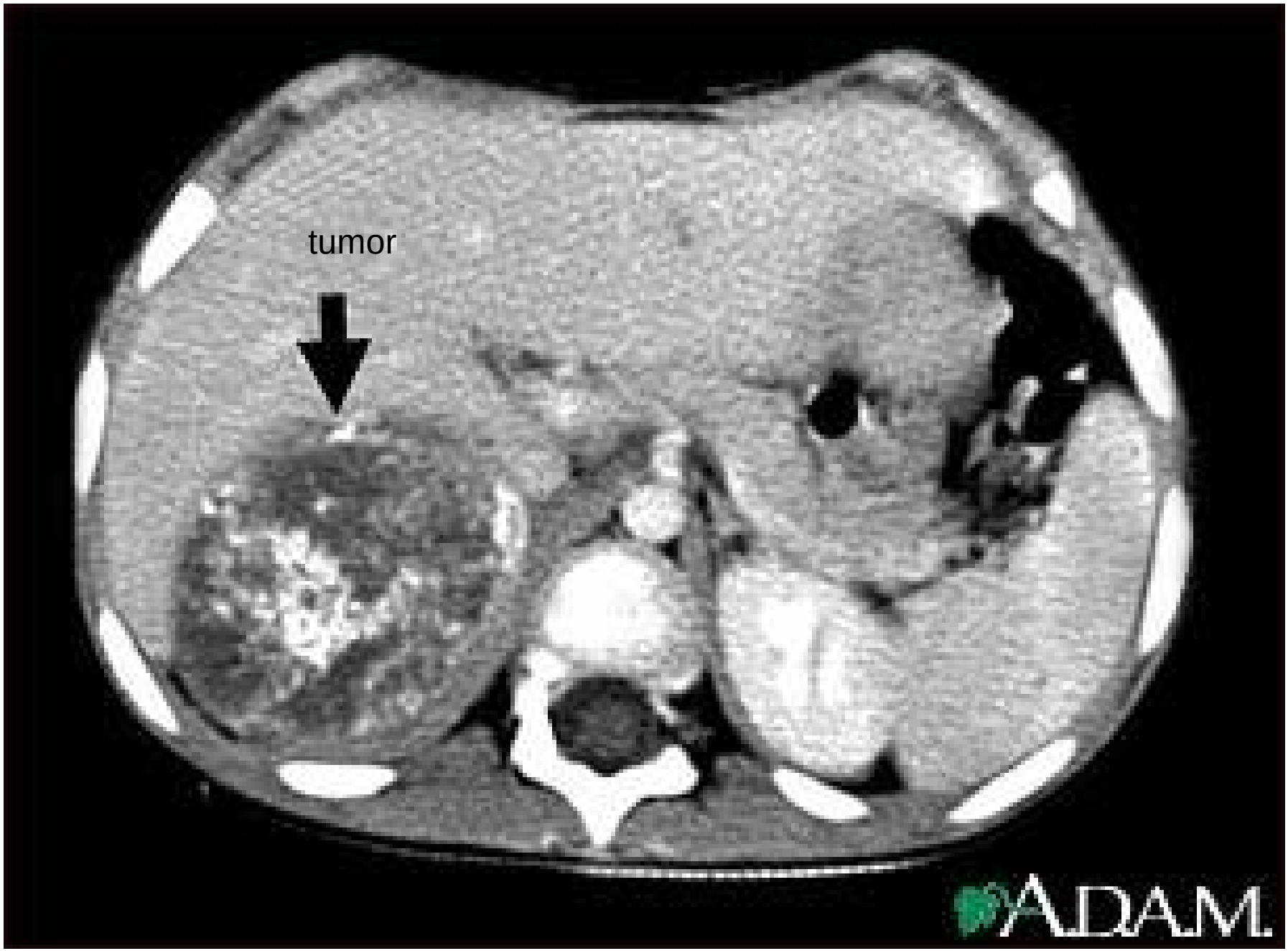
- *CBC : decrease Hb , plat. ,& orWBC. May indicate bone marrow involvement.
- *Urine Catecholamines: elevated Homovanillic acid (HVA) Vanillylmandelic acid(VMA) in 90% of patient.
- *Liver function test :may indicate liver involvement.
- *Lactate dehydrogenase:have prognostic value.
- *Bone marrow aspirate&biopsy: to evaluate bone marrow involvement.
- X-ray

Diagnosis

- *Plain film of primary site ,intravenous pyelogram
- *Us, CT,and or MRI of primary site.
- *Skeletal survey or bone scan to rule out bone lesion.
- Definitive diagnosis:histological characteristics on diagnostic biopsy.
- Staging
- Low _risk group; usually have localized disease.
- High _risk group:disseminated disease often involve the bone,bone marrow, liver and/or skin.

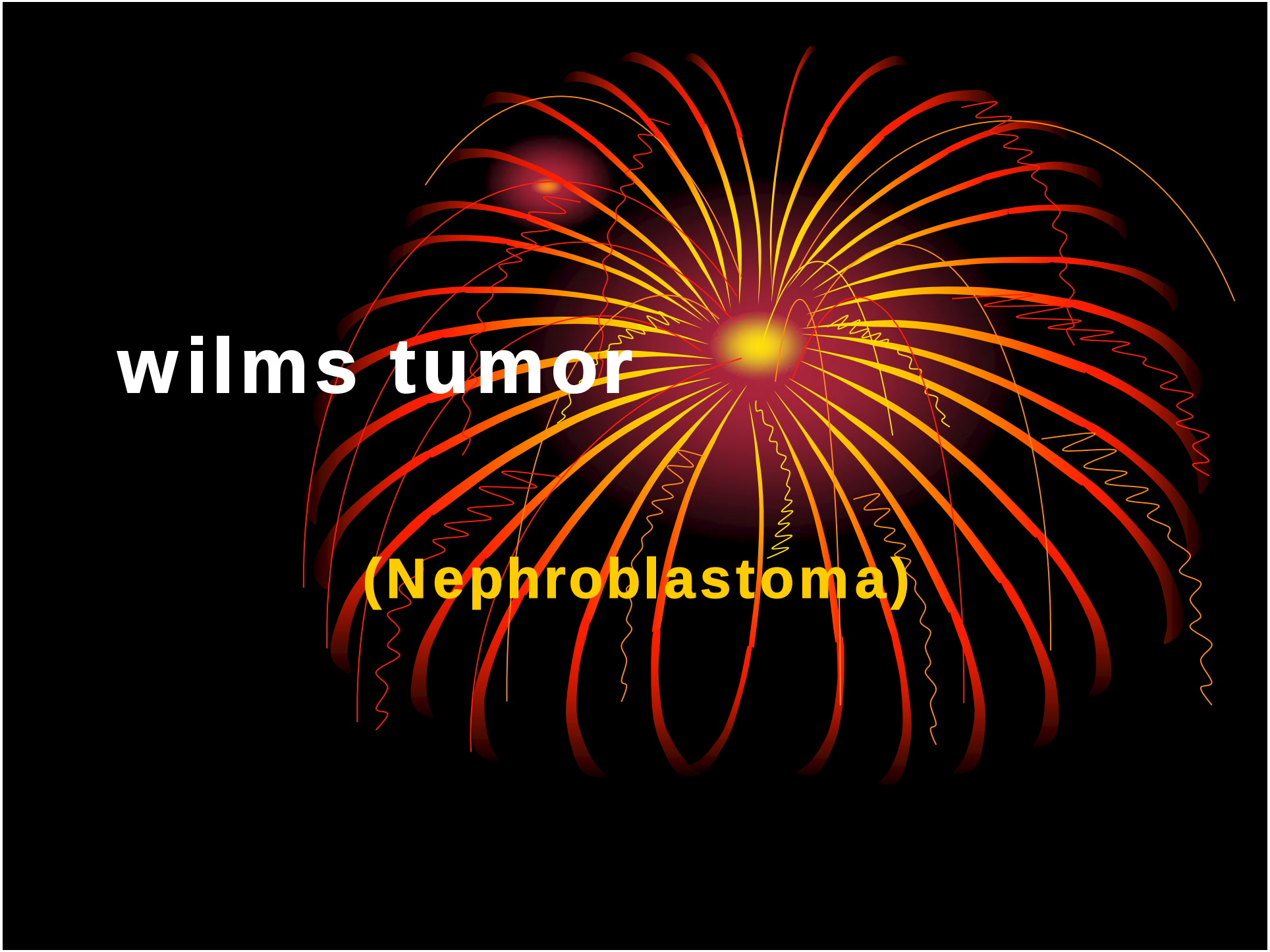
calcification

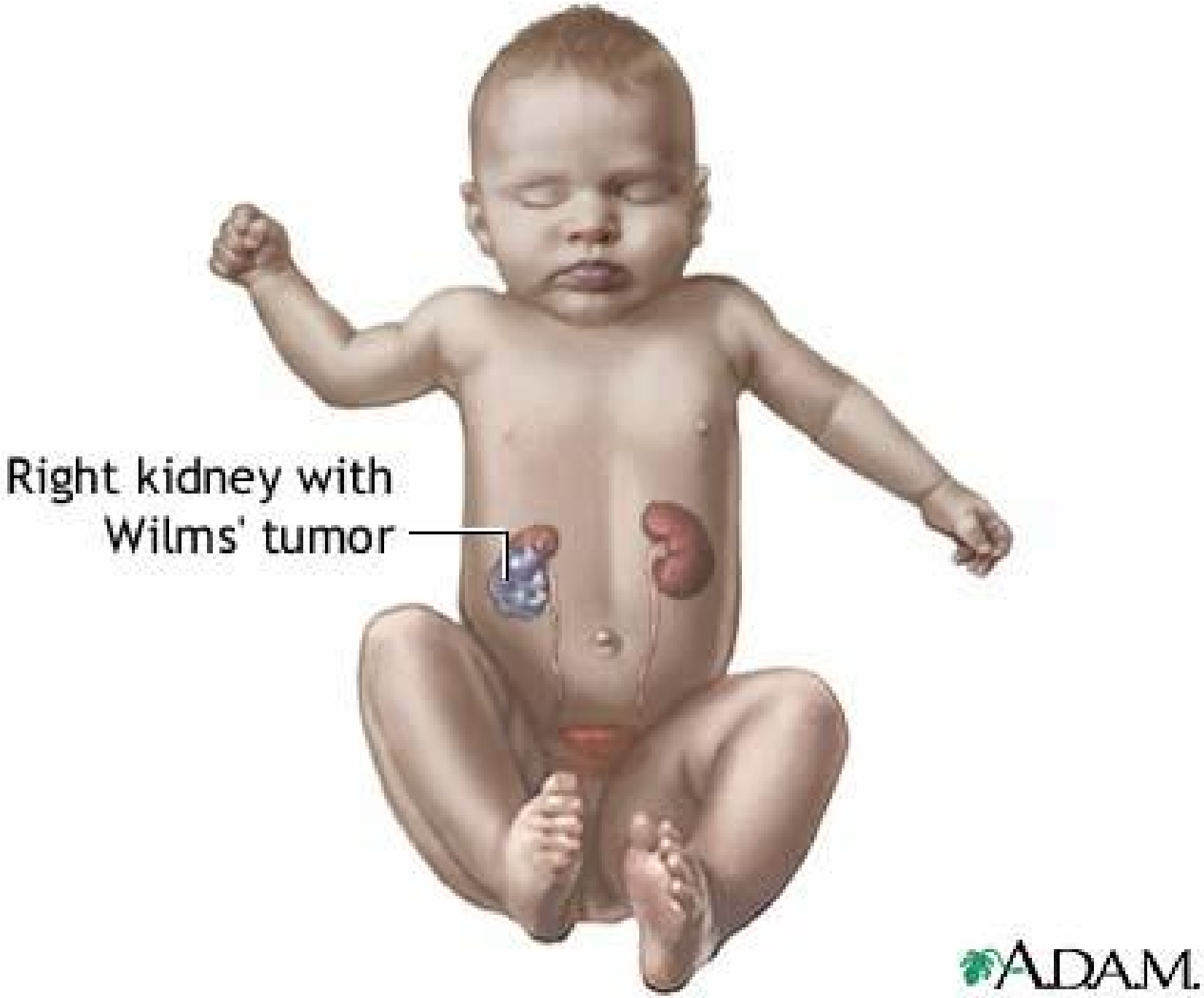




Treatment

- *Surgical excision : is best for localized disease.
- *Chemotherapy ;(with cyclophosphamid & doxorubicine) can induce remission in 50% of patient
- * Local radiation can cause regression of tumor & is used to treat symptom (pain)
- *BM transplantation ; used in several protocol s
- prognosis
- is good with young age& localized tumor





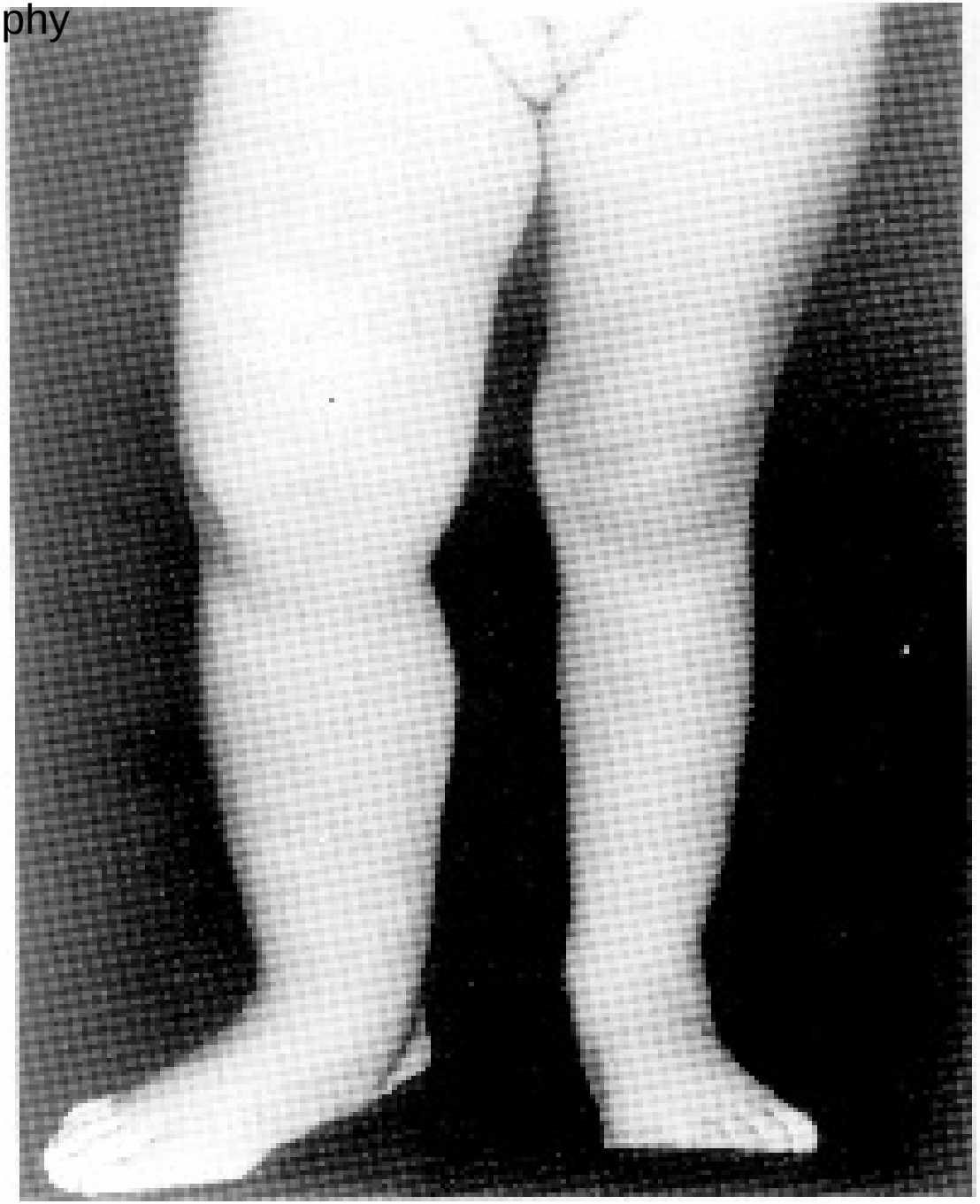
wilms tumor (Nephroblastoma)

- Malignant tumor of the kidney ,occurring in the pediatric age group

Causes; (unknown)

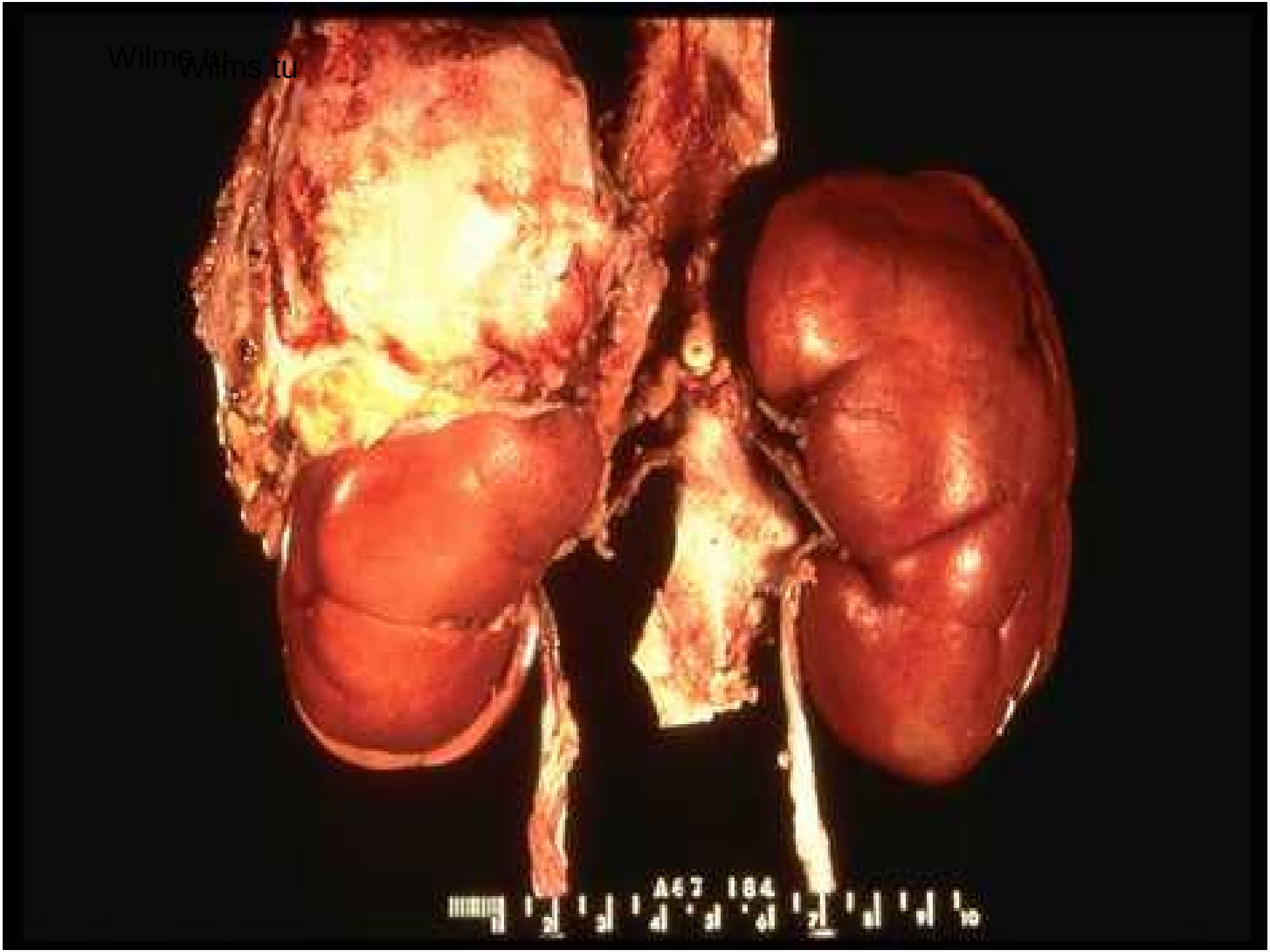
- *5-6% of all childhood cancer
- * peak age 2-3 years
- * may be associated with aniridia ,hemihypertophy ,cryptorchidism
- *Familial cases are more often bilateral and occur at younger age .
- *Tumor suppressor gene cloned at chromosome 11p 13.

hemihypertrophy



Pathology

- Wilms tumor is solitary growth ,cystic with hemorrhage usually no calcification (differ from Neuroblastoma)
- Histology ; triphasic pattern ; blastemal , epithelial ,&stromal cell
- Presence of anaplasia indicate poor prognosis



Staging ;(NWTS) national wilms tumor staging

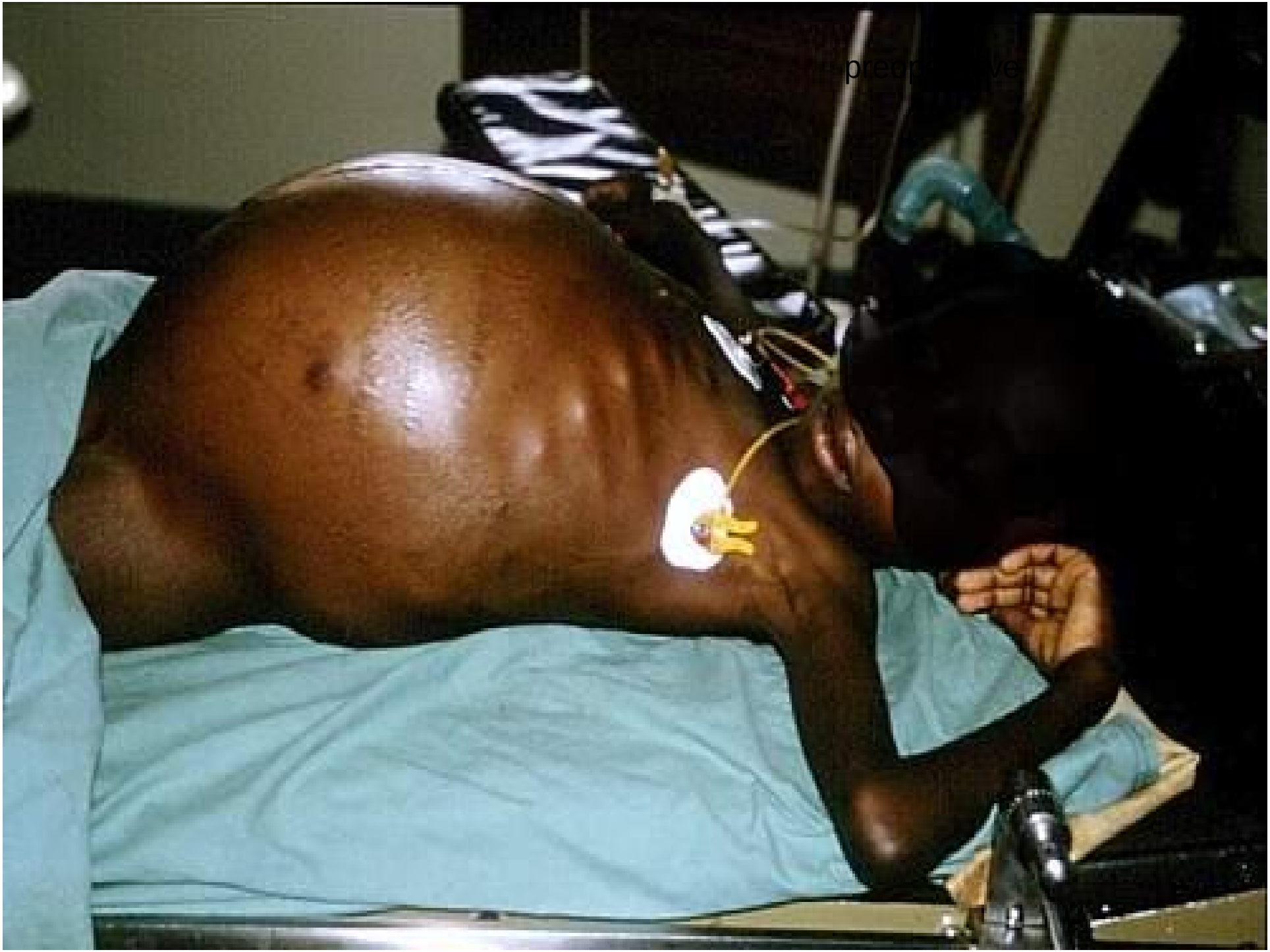
- *Stage I ;tumor restricted to one kidney &completely resected without rupture of capsule
- *Stage II ; tumor extend beyond capsule ,but is resected totally .
- * Stage III; residual non hematogenous tumor confined to abdomen
- *Stage IV; hematogenous spread to lung & liver
- * Stage V; bilateral disease

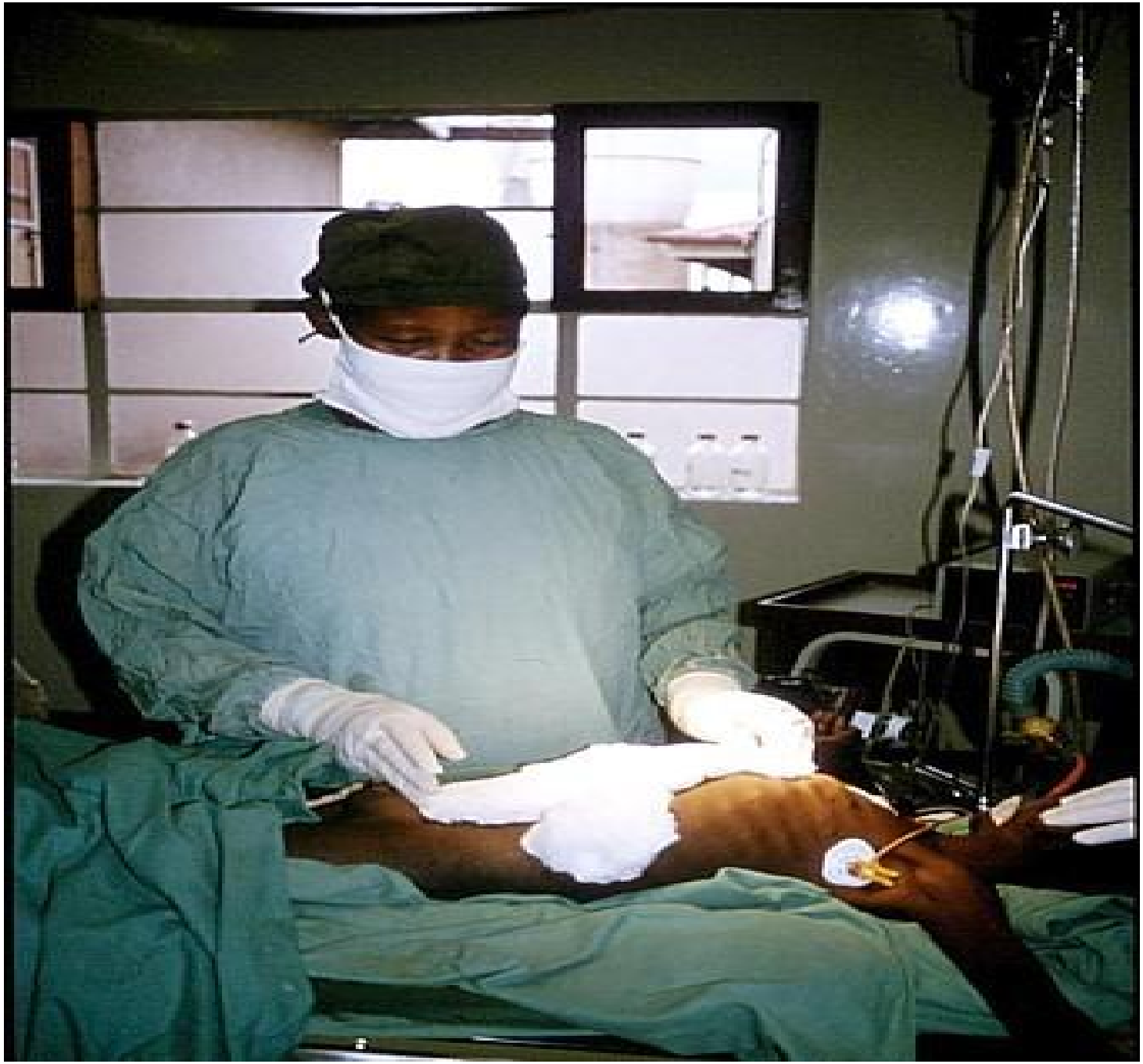
Differential diagnosis

- 1-polycystic kidney
- 2- renal hematoma
- 3- renal abscess
- 4-neuroblastoma
- 5- other neoplasm of the kidney (clear cell carcinoma)

Clinical manifestation

- abdominal distention ; firm abdominal mass that doesn’t cross midline
- abdominal pain or vomiting .
- hematuria; 25%
- hypertention;resulting from pressure on renal artery
- rapid increase in abdominal size (suggestive of hemorrhage in the tumor)
- fever r
- anemia ;secondary to hemorrhage in the tumor





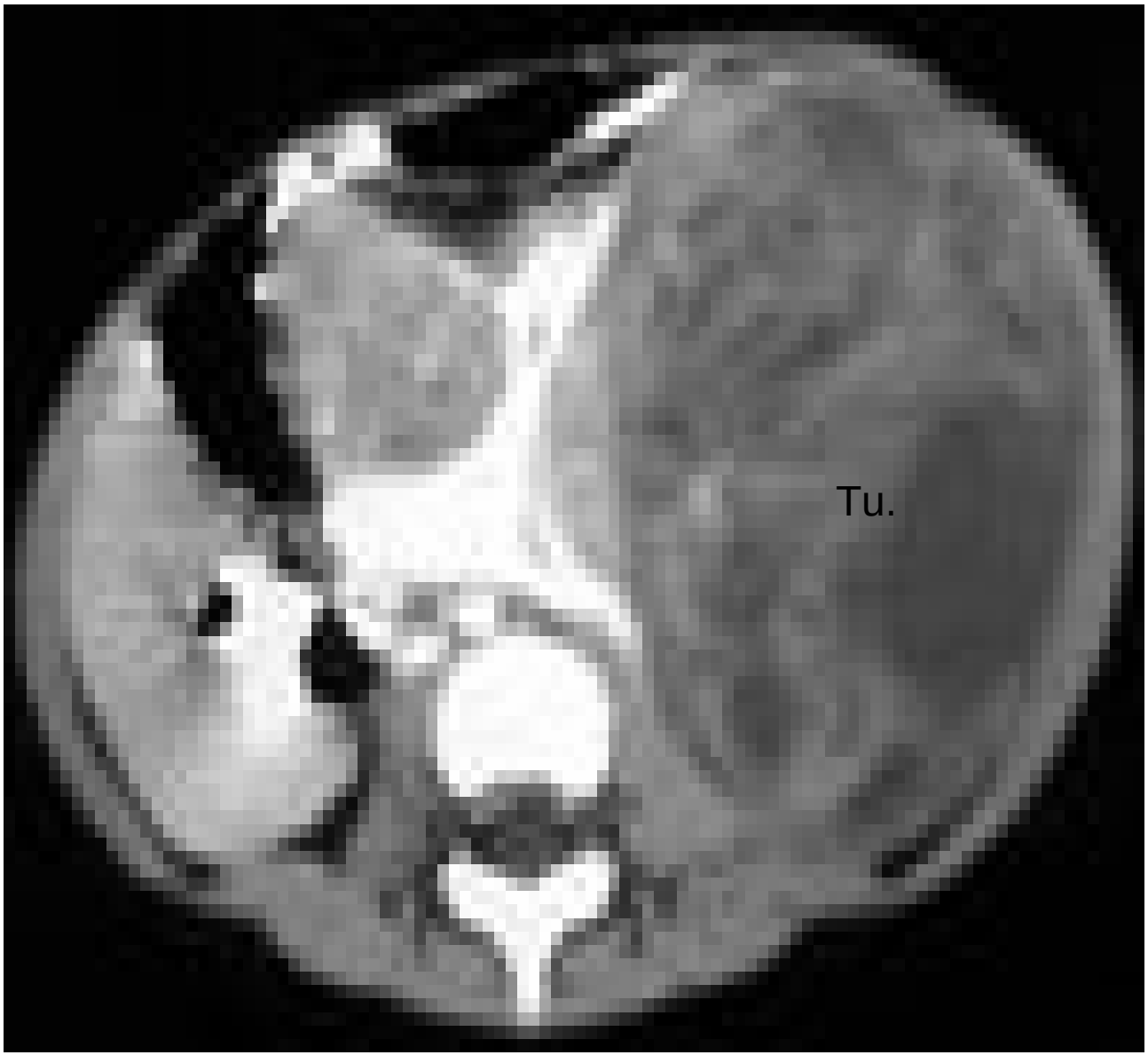
Post operative

Diagnosis

- CBC
- Urine analysis ; show hematuria
- Liver & kidney function test
- US of the abdomen (intrarenal mass)

Diagnosis

- CT scan ,show inhomogeneous mass with area of low density indicate necrosis, calcification are less common than Neuroblastoma,also it determine the intrarenal source of tumor &the extent and evaluate the opposite kidney
- CXR ;evaluate for metastatic disease
- Bone scan & bone marrow ; only in unfavorable pathology



CT-scane

Therapy

- Surgery ;nephrectomy
- Radiation ; not required for stage I ,II
- Local X-Ray therapy for stage III,IV
- *Chemotherapy ;stage I, II, -----every 3 weeks for 6 months
- stage III,IV --- for 6-15 months.

Prognosis

- Stage I,II greater than 90% cured
- Stage III 85 % cured
- Stage IV 70 % cured