

History

- Eyad is 3 year old saudi boy brought to ER by his parents with history of
- abdominal pain ,
- constipation and
- weight loss ,
- Rapid breathing since one week.

AL.BAHA GRAND ROUND

Prepared by
Pediatric department

Cont. history

Past medical history:

- He is product of full term ,SVD , no neonatal admission
- No history of previous admission.
- No history of similar complaint.
- No history any medical or surgical illness.
- Remaining history is unremarkable.

-No history of vomiting or urinary symptoms.

-No history of respiratory symptoms.

-No history of loss of consciousness or abnormal movement.

-No history of skin rash.

Examination

- Chest : tachypnic , subcostal& intercostal retraction, with crackles and bronchial breathing mainly in left side of chest.

Examination

- Patient is conscious ,alert ,febrile, not pale, jaundiced,or cyanosed but distressed , fairly hydrated & perfused, no dysmorphic features
- Vital signs:
 - T:38.2c , HR; 126 , RR:45 BP:150/84 mmHg.
 - O2 sat.93% RA.
 - HEENT: clinically free.
 - Eye examination normal.

Examination

- Liver and spleen are not palpable.
- No lymph node palpable and joint examination normal.
- Intact hernial orifices.
- CNS: conscious ,hypoactive ,normal tone, power and reflexes ,intact cranial nerves and cerebellar function.
- Normal male genitalia.

Abdomen :

distended abdomen , visible bowel loops in upper abdomen
 Palpable mass in right hypochondrium , not tender 8x9 cm , smooth surface, firm in consistency ,fixed , ill-defined edge mass is ballotable.

Investigations

- CBC : WBC 10,000 , Hb: 10.8 g/dl. Plt.227,000
- Diff: neutrophil 54% , lymphocytes 44%
- ESR : 20 mm/h
- Peripheral smear : normal.
- Urea and electrolytes normal.
- LFT : all within normal except LDH 567.
- Urine analysis: pus cells 20-30 cell/hpf

Differential diagnosis

1. Wilms tumor .
2. Neuroblastoma.
3. Hydronephrosis.
4. Non-Hodjkin lymphoma.



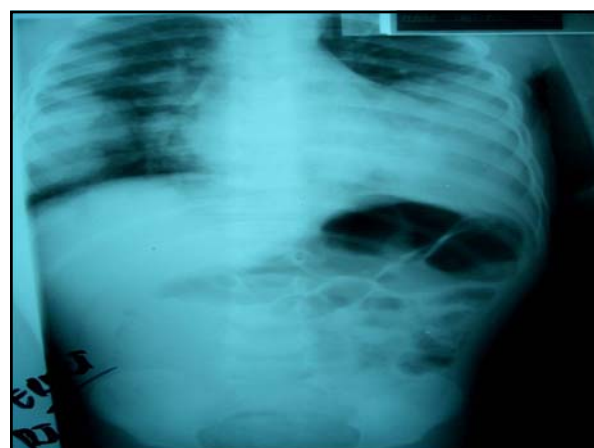
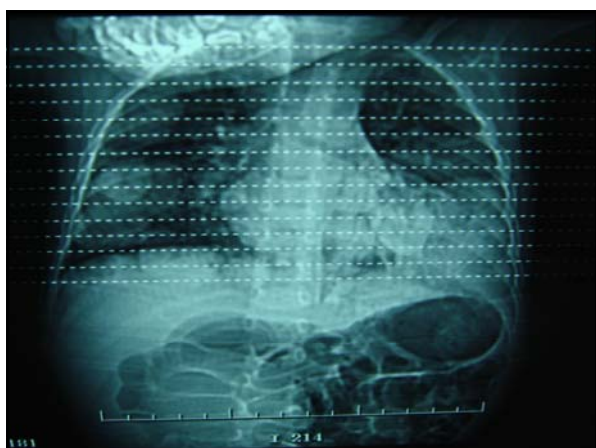
Investigations

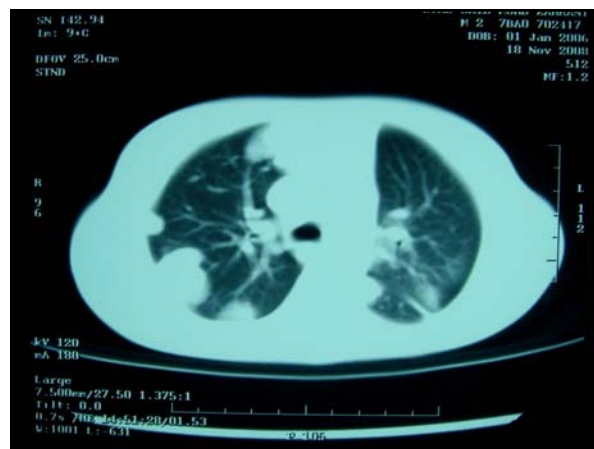
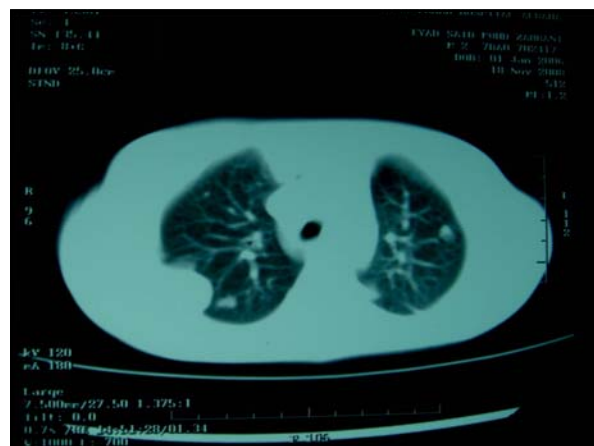
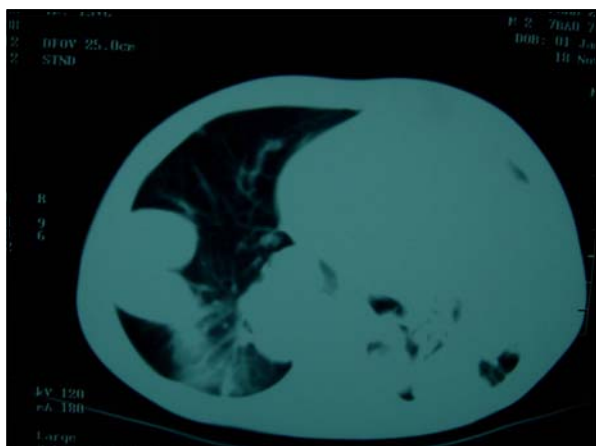
- Ultrasound abdomen : mass 8x9cm at site of right kidney , no kidney tissue can be detected.
- CT abdomen : retroperitoneal mass with calcification and total destruction of right kidney.



Investigations

- CXR: multiple round opacities and left pleural effusion, ? Metastasis.
- CT chest: chest multiple metastasis with evidence of calcification within the lung.





Final diagnosis

- Stage IV wilms tumor

Investigations

- VMA , HVA, and CEA not available.
- Skeletal survey normal.

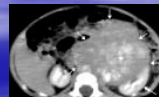
Wilms Tumor

- Pathology
 - large, bulky, well-encapsulated lesions
 - propensity for venous extension in renal vein, IVC, RA
 - histology is tri-phasic: blastemal, stroma, and epithelial elements
 - FH vs. UH (anaplastic) histology affects prognosis

Pearls

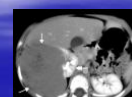
Neuroblastoma:

- irritable child, tender
- skin: blueberry muffin
- eyes: raccoon eyes
- some degree of wasting
- urinary metabolites
- calcs on film



Wilms' Tumor:

- asymptomatic
- macroglossia
- aniridia
- hemihypertrophy
- "claw" on CT/IVP
- hypertension
- hematuria



Wilms' Tumor



Wilms' Tumor

- Pre-treatment imaging
 - CXR, AXR (linear calcifications)
 - USG of kidney and venous drainage
 - CT scan of abdomen +/- chest

Stages of Wilms tumor

- Stage III: residual nonhematogenous tumor to the abdomen. Lymph node involvement hilus, periaortic chain, or beyond; diffuse peritoneal contamination by tumor spillage; peritoneal implants of tumor; tumor extends beyond surgical margins microscopically or macroscopically; tumor not completely removable because of local infiltration into vital structures.

Stages of Wilms tumor

- Stage I : tumor limited to kidney and completely excised. Capsular surface intact ; no tumor rupture; no residual tumor apparent beyond margins of excision.
- Stage II : tumor extends beyond kidney but is completely excised. Regional extension of tumor, vessel infiltration; tumor biopsied or local spillage of tumor confined to the flank. No residual tumor apparent at or beyond margins of excision.

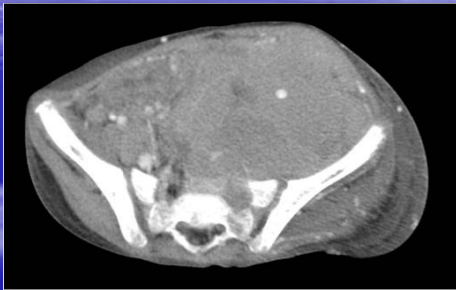
Neuroblastoma

- Most commonly presents with abdominal mass
 - constitutional symptoms: fever, weight loss, anemia, FTT, bone pain
- Metastases at presentation in 3/4 of patients
 - bone, BM, and lymph nodes most common
 - liver and skin less frequently, rare lung and brain

Stages of Wilms tumor

- Stage IV: deposits beyond stage III (e.g., lung, liver, bone, brain)
- Stage V: bilateral renal involvement at diagnosis.

Neuroblastoma



Neuroblastoma

- X-rays may reveal stippled calcifications
- Pre-treatment staging essential
 - CT scan, MIBG scan, BM biopsy, urine catecholamines

Stages of neuroblastoma

- Stage I: Tumors confined to organ or structure of origin .
- Stage II: tumors extends beyond the structure of origin but not across the midline, with(stage 2B) or without (stage 2A) ipsilateral lymph node involvement.
- Stage III: tumors extend beyond the midline, with or without bilateral lymph node involvement.

Neuroblastoma



Cont. neuroblastoma

- Prognosis depends on age, stage, histology (Shimada classification), and genetic factors
 - poor prognosis with *N-myc* amplification, allelic loss of 1p, MDR over-expression, normal ploidy
- Staging by INSS depends on localization and excision
- Survival is improving
 - stage I 90% 4-yr survival
 - stage IV 15-40% 4-yr survival after BM transplant

Stages of neuroblastoma

- Stage IV: tumors disseminated to distant sites(e.g., bone, bone marrow, liver, distant lymph nodes, other organs).
- Stage IVS: refers to children younger than 1 year of age with dissemination to liver, skin , or bone marrow without bone involvement and with a primary tumor that would otherwise be stage I or II.

