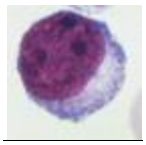
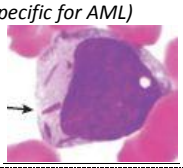
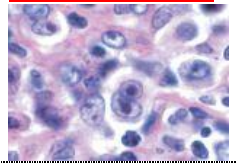
# Oncology Notes

## Leukemia

| | Acute Lymphoblastic Leukemia | Acute Myelocytic Leukemia |
|---|---|---|
| **Cells of Origin** | **- Predominant cells are of *Lymphoid origin***<br>**- Most ALL originates in *B-Cell Precursors***<br>- **Philadelphia chromosome**<br>t (9:22) or *BCR-ABL* genes<br>* is almost always seen in **CML**<br>* BUT may also be seen in about 5% of ALL | **- Predominant cells are of *myeloid origin*** |
| **Clinical Features** | **Child from 2 – 10 years , with:**<br>1- Exertional Dyspnea / Pallor<br>2- Easy bruising / purpura<br>3- **Infections:**<br>- High grade fever<br>- Otitis Media<br>- Sore throat<br>4- **Lymphadenopathy**<br>5- **HepatoSplenomegaly** | - Both → **Children & Adult affected** :<br>1- Exertional Dyspnea / Pallor<br>2- Easy bruising / purpura<br>3- **Infections:**<br>- High grade fever<br>- Otitis Media<br>- Sore throat<br>5- **HepatoSplenomegaly**<br>6- Mucosal Bleeding & Ocular hemorrhage |
| **Lab** | - Anemia - Thrombocytopenia - Neutropenia | - Anemia - Thrombocytopenia - Neutropenia |
| **Peripheral Smear** | - **Lymphoblasts** (Large nuclei & Prominent nucleoli)<br>  | 1- Large myeloblasts with **notched nuclei .**<br>2- and **Auer Rods** *(specific for AML)*<br>  |
| **Bone Marrow** | Diagnostic: **>25% lymphoblasts** | |
| **Histochemical Staining** | **Positive "Periodic Acid Schiff (PAS)" reaction**<br>*Lymphoblasts contain cytoplasmic" PAS"materials*<br>*BUT lack "perioxidase positive" granules* | **Myeloblasts** has : **"perioxidase positive" granules**<br>→ staining with **myeloperoxidase** |
| **ImmunoStaining** | **For terminal deoxynucleotidyltransferase (TdT) is Positive** (TdT expressed only by Pre-T/Pre-B lymphoblasts) | |

## Lymphoma

| | Hodgkin Lymphoma | Non-Hodgkin Lymphoma |
|---|---|---|
| **Cells of origin** | **B cells** | 1- Lymphocytes (**most commonly B cells**) or<br>2- Natural killer cells |
| **Classification (low to higher grade)** | **1- Nodular sclerosis** (**most common**, women = men, fibrosis of lymph nodes)<br>**2- Mixed cellularity**<br>**3- Lymphocyte Rich** (rare, **best** prognosis)<br>**4- Lymphocyte-depleted** (very rare, **worst** prognosis) | Many types, Common variants include:<br>1- Diffuse large B cell (**most common**)<br>2- Follicular small cell **(B cells, t[14;18])**<br>3- Small lymphocytic **(same disease as CLL)**<br>4- **Burkitt ( EBV related**, t[8;14], **"starry sky" pattern)**<br>5- Peripheral (**T cells**) |
| **Risk factors, patient population** | - **Bimodal age distribution** (peaks at 20 and 65 yr of age)<br>- **men > women** (except for nodular sclerosis subtype) | 1- EBV → **Burkitt Lymphoma** :<br>- Most patients present with either:<br>1- Mass involving Mandible or<br>2- Abdominal Viscera<br>2- HIV<br>3- Congenital immunodeficiencies<br>4- Rheumatic disease |
| **Clinical Features** | 1- Painless lymphadenopathy **(neck)**<br>**2- Fever , night sweats , weight loss,**<br>3- Pruritus,<br>4- Hepatosplenomegaly | 1- Painless lymphadenopathy **(generalized)**<br>**2- weight loss, fever, night sweats** |
| **Labs** | LN biopsy shows : **Reed-Sternberg cells** **(resemble owls' eyes)**<br>  | **Lymph node or bone marrow biopsy shows :**<br>**Lymphocyte proliferation**<br>(**cleaved cells seen in follicular small cell variant**) |
| **Treatment** | **Radiation, chemotherapy** | **Palliative radiation, chemotherapy** |
| **Prognosis** | **Good**, 80% cure rate unless far progressed | **Poor** (months for aggressive types, years for less aggressive variants), worsens with increasing age |

# Brain Tumors

- In pediatrics, **CNS tumors** are the **most common solid tumors** and the **second most common malignancies** (after leukemias).
- **Astrocytomas are the most common histologic type** for both supratentorial & infratentorial groups

- **Anterior vermis lesions** are due to degeneration from **Alcohol abuse** and present with **GAIT ATAXIA**
- **Posterior vermis lesions** are due to **Medulloblastoma or Ependymoma** and present with **TRUNCAL ATAXIA**

| Age | Location 15% arise in midline | Types |
|---|---|---|
| **0-1 year** | Supra-tentorial (25% ) | 1- Choroid plexus tumor<br>2- Teratoma |
| **2-10 year** | Infratentorial (60% ) | 1- Juvenile pilocytic astrocytoma<br>2- Medulloblastoma |
| **>10 years** | Supratentorial | 1- Diffuse astrocytoma |

## Medulloblastoma

- **Embryonal tumor** arising from **external granular layer of cerebellum in cerebellar vermis (esp posterior)**
- **Site**: Most common in **Posterior vermis in midline** (2nd most common tumor of posterior fossa in children)
- May **obstruct 4th ventricle giving hydrocephalus**
- **Presents with**: **1- Unbalanced gait** **2- Truncal ataxia** **3- Horizontal nystagmus** (Posterior vermis syndrome)
- **Treatment:** with **Radiation & chemotherapy** (it is highly aggressive but responds to chemo)
  - Can give drop metastasis
  - Highly radiosensitive

## Craniopharyngiomas

**Cystic Calcified parasellar lesion on MRI**

- Craniopharyngiomas are derived from **epithelial remnants of Rathke's pouch**
- It is not a true pituitary tumor, and is characterized by **nests of squamous cells in a loose stroma**, resembling the **appearance of embryonic tooth bud enamel**
- **A young boy** with symptoms of ***:***
  1- ***Increased intra-cranial pressure*** (e.g .. headaches vomiting)
  2- ***Bi-temporal hemianopsia*** (because Its location is suprasellar and inferior to the optic chiasm)
  3- Calcified lesion above the sella
  → has a **craniopharyngioma** until proven otherwise.
- **Associations :**
  1- **Endocrine symptoms** such as ***diabetes insipidus***, and ***growth failure*** associated with either ***hypothyroidism*** *or* ***growth hormone deficiency***.
- **Diagnosis :** Usually made by (***CT***) or (***MR***) imaging:
  - Presence of a **Cystic Calcified parasellar lesion on MRI** is almost diagnostic of craniopharyngioma.
- **Treatment :** Surgical removal is the treatment of choice.

## Pinealoma

- Develops in the **dorsal aspect of the midbrain**
- **Symptoms** consist of ***:*** 1- **Endocrine syndrome** 2- **Intracranial hypertension**
  **3- Parinaud's sign** (paralysis of vertical gaze) 4- **Collier's sign** (retraction of eyelid).

# Painless abdominal mass in a child <5 years

| Wilms Tumor (Nephroblastoma) | Neuroblastoma |
|---|---|
| Arise from **Kidney (metanephros)** | Arise from **Neural crest cells** of **sympathetic nervous system** (similar to pheochromocytoma but fewer cardiac findings) |
| - Most common primary renal neoplasm of childhood<br>- Usually diagnosed between the ages of **2 – 5 years** | - Most common extracranial solid cancer in childhood and the most common cancer in **infancy <1 year** |
| 1- Most cases are sporadic<br>2- Mutations of **WT1 gene on chromosome 11p13** | **N-myc proto-oncogene** |
| **- Asyptomatic non tender smooth abdominal mass** ( in 80% )<br>- **The mass bilateral** ( in 10% of cases ) | Most common site: **Adrenal, Retroperitoneal ganglion**<br>**Non tender abdominal, thoracic, cervical mass** |
| **Symptoms:**<br>1- Hypertension<br>2- Hematuria<br>3- Abdominal Pain & vomiting<br>4- Lung metastasis (in few cases) | **Symptoms:**<br>1- Can cause hypertension<br>2- **Mostly metastatic at the time of diagnosis**: long bones, skull, bone marrow, liver, lymph nodes |
| **Kidneys are separately palpable from tumor** | **Kidneys are non-palpable** |
| **Don't cross the midline** | **Cross the midline** |
| **Associated with:**<br>1- **WAGR Syndrome: W**ilms **A**niridia **G**U anomalies **M**ental Retardation<br>2- **Beckwith widemann syndrome**<br>3- **Denys-Drash syndromes**<br>4- H/O **Horseshoe kidney** | **Associated with:**<br>1- **Hypsarrhythmia** (dancing eyes)<br>2- **Opsoclonous** (dancing feet)<br>**3- Horner syndrome** |
| **Best initial test** = **USG** | **Elevated 24 hours urinary catecholamines** (VMA & HVA) |
| **Most accurate** = **CECT** | **Calcifications on imaging** |
| **Treatment**: is **nephrectomy**.<br>If treated in the early stage, majority of the patients have a long-term survival. | |