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Introduction

Leukostasis

- Leukostasis comes with Hyperleukocytosis (Total leukocytes > 50*10^9/L)
- Medical emergency!!!
- **Seen in pts with AML or CML in blast crisis** (more common in myeloid blasts, most common AML)
  - 10-20% in newly diagnosed AML (WBC > 100)
  - 10-20% in newly diagnosed ALL
  - Rare in CLL (except if WBC > 400 - cells are too small to occlude)
  - Rare in CML (unless myeloid blast crisis with high blast counts, WBC > 250)
- Extremely elevated blast cell count and symptoms of decreased tissue perfusion

Diagnosis of Leukostasis

- **Diagnosis is Clinical**
- Features:
  - 1. Presence of Leukemia
  - 2. Hyperleukocytosis (leusks > 50*10^9/L)
  - 3. Respiratory or neurological distress

- Mortality --> 20-40%
- Symptoms:

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Source: Created from Dr. Yulia Lin’s slides
<table>
<thead>
<tr>
<th>Organ</th>
<th>Manifestations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary</td>
<td>Dyspnea, Hypoxemia (+/- interstitial/alveolar infiltrates)</td>
</tr>
<tr>
<td>Neurological</td>
<td>Visual Changes, Headache, Dizziness, Tinnitus, Gait Instability, Confusion, Somnolence, Coma</td>
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<tr>
<td>Infection</td>
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<td>Cardiac</td>
<td>Myocardial ischemia</td>
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<td>Renal</td>
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<td>MSK</td>
<td>Acute limb ischemia</td>
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<td>GI</td>
<td>Bowel Infarction</td>
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</tbody>
</table>

- **Investigations:**
  - Platelets (overestimated --> blast fragments misread as platelets)
  - Hyperkalemia
  - DIC (decreased fibrinogen, increased D-dimers)
  - Spontaneous Tumor Lysis Syndrome (TLS)
    - Elevated uric acid, hyperkalemia, hyperphosphatemia, hypocalcemia

- **Management**
  - Stabilize + Lower WBC count!
  - Cytoreduction
    - Chemotherapy or leukapheresis (often induction chemotherapy, or rarely hydroxyurea)
  - **TLS Prophylaxis**
    - Aggressive Hydration!
    - Allopurinol
  - Supportive Care
    - Monitor for DIC
    - Platelet transfusions (keep > 20-30,000 /μL)
    - Avoid blood transfusion (can increase viscosity)

### Hyperviscosity Syndrome

- Elevated blood plasma viscosity causing neurological signs (vision loss, headache, tinnitus, ataxia)

- **Occurs in**
  - Waldenstrom's macroglobulinemia (large IgM pentamers)
  - Multiple Myeloma
  - Rheumatoid disease
  - Polycythemia
  - Sickle Cell Disease
  - Leukemia
  - Spherocytosis

- **Symptoms**
  - **Organ** | **Symptoms**
<p>| | |
|---|---|
| Hematologic | Mucosal bleeding |</p>
<table>
<thead>
<tr>
<th>Organ</th>
<th>Symptoms</th>
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<tbody>
<tr>
<td>Abnormal bloodwork</td>
<td></td>
</tr>
<tr>
<td>Neurologic</td>
<td>Headache, vision loss, vertigo, nystagmus,</td>
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<td></td>
<td>tinnitus, deafness, diplopia</td>
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<tr>
<td>Fundoscopic Examination</td>
<td>Dilated segmented tortuous retinal veins</td>
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<tr>
<td>(Very important!!!)</td>
<td>Hemorrhage</td>
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<tr>
<td></td>
<td>Papilledema</td>
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<tr>
<td></td>
<td>Exudates</td>
</tr>
<tr>
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<td>Central retinal vein thrombosis</td>
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**Organ** | **Symptoms** |
--- | --- |
Abnormal bloodwork |  
Headache, vision loss, vertigo, nystagmus, tinnitus, deafness, diplopia |
Dilated segmented tortuous retinal veins |  
Hemorrhage |
Papilledema |  
Exudates |
Central retinal vein thrombosis |

- **Investigations**
  - Viscosity (normal level 1.4-1.8)
    - Hyperviscosity unlikely with viscosity < 4
- **Management**
  - Plasmapheresis + Chemotherapy
    - Plasmapheresis --> reverses retinopathy +sx (reduces viscosity by 20-30%/session)
    - Chemotherapy --> Start with plasmapheresis
  - Repeat retinal examination

### Tumor Lysis Syndrome

- Spontaneous or chemo-induced release of intracellular electrolytes & nucleic acids
- **Most common:**
  - High-Grade Burkitt's Lymphoma
  - Leukemias (ALL, AML, CML in blast crisis)
  - (solid tumors rarely)
- **Clinical Features**
  - High K+
  - High Uric Acid
  - High PO4
  - Low Ca++
  - Renal Failure (urate nephropathy)
- **Prophylaxis**
  - Allopurinol 300mg qd to BID PO (or 200-400mg/m² 1IV adjusted for renal function)
    - + Aggressive hydration prior to chemo
  - OR Rasburicase (urate oxidase) 0.15 mg/kg or 6mg fixed dose (except in obese pts) + hydration
- **Treatment**
  - Avoid IV contrast & NSAIDs
  - **Allopurinol + aggressive IV fluids + diuretics** (to U/O of 80-100cc/hr)
    - Consider alkalination of urine w/ isotonic NaHCO3 (inrr urine urate excretion, reduces nephropathy, but controversial (avoid with rasburicase risk of met alkalosis or CaPO4 precip)
  - Rasburicase 0.1-0.2 mg/kg x1 repeat as indicated (consider, if very high uric acid levels and aggressive malignancy) contraindicated in G6PD def (causes hemolytic anemia)
    - UA must be drawn on ice (otherwise rasburicase removes it in vitro)
  - Treat HyperK+, HyperPO4-, HypoCa++
  - Consider IHD if severe AKI