Acute Promyelocytic Leukemia (APL) Clinical Presentation

- Younger patients—median age 47 years
- Sudden Onset
  - Fatigue
  - Petechiae, ecchymosis
  - Mucosal or retinal hemorrhage
- Typically pancytopenia on lab tests

APL Unique Features

- t(15;17) producing PML-RARA fusion protein
  - Abnormal function of the retinoic acid receptor
  - Disrupted differentiation of myeloid progenitors
  - Promyelocytes and blasts increase
- Propensity for DIC
  - Increased expression of procoagulants
  - Enhanced fibrinolysis

Transfusion Indications

- RBCs
  - Deficit of O₂ carrying capacity
  - Hgb <7 g/dL for most people
- Plasma
  - Deficiency of multiple coagulation factors: liver disease, warfarin therapy, massive transfusion, DIC
  - Specific factor/protein deficiency, no concentrate
  - INR >1.5 (1.8?)


Curr Opin Hematol. 2016 Mar;23(2):121-6

Transfusion Indications

- **Platelets**
  - Prophylaxis
    - Non-bleeding patients
    - Platelet count <10x10⁹/L
  - Treatment
    - Bleeding/surgical patient
    - Platelet count <50x10⁹/L typically
    - Neurological often <100x10⁹/L
    - Platelet dysfunction (aspirin, Plavix, urea, plastic)

- **Cryoprecipitated AHF**
  - Fibrinogen <100 mg/dL


Blood Product Special Needs

- **Leukocyte reduction**
  - Reduce febrile non-hemolytic transfusion reactions
  - Decrease HLA alloimmunization
  - Decrease CMV transmission

- **Irradiation**
  - Prevent transfusion associated GVHD

- Possibly CMV screening for potential HCT candidates

NCCN Guidelines Version 1.2015

Medical Emergency

- 17-29% of patients die in first 30 days
- Severe hemorrhage
  - Intracranial
  - Pulmonary
- Differentiation syndrome
- Infection

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NCCN Guidelines Version 1.2015
APL Treatment

- High risk WBC >10,000/µL
- ATRA +/- Chemotherapy +/- ATO
  - ATRA targets RARA
  - ATO targets PML
- Degradation of PML/RARA fusion protein
  - Maturation of leukemic cells
  - Resolution of coagulopathy

Br J Haematol. 2015 Dec 21

Supportive Measures

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<th>Lab Parameter</th>
<th>NCCN APL</th>
<th>ELN APL</th>
<th>NCCN &amp; ELN AML</th>
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<td>Hemoglobin (g/dL)</td>
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<td>&gt;8</td>
<td>&gt;8</td>
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<td>Platelet Count (x10^9/L)</td>
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<td>&gt;30-50</td>
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<tr>
<td>PTT (sec)</td>
<td>Close to normal</td>
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</table>

European LeukemiaNet. Blood. 2009 Feb 26;113(9):1875-91

At the Bedside

- Clerical check
- Visual check
- 170-260µ filter removes fibrin clots, aggregates
- 22-14 gauge needle/catheter
- 0.9% (normal) saline
- Appropriate blood warmers
- Pre-medication
- Transfusion must be completed within 4 hrs
- Stop transfusion if suspected reaction

Types of Transfusion Reactions

- Fever
  - Febrile
  - Hemolytic (delayed vs. acute)
  - Bacterial sepsis
- Respiratory distress
  - Transfusion related acute lung injury (TRALI)
  - Transfusion associated circulatory overload (TACO)
  - Allergic (anaphylaxis)
- Rash
  - Allergic
  - Thrombocytopenia
    - -- Postransfusion purpura
    - TA-GVHD
      -- Platelet refractory


Bibliography