

MYELOYDYSPLASTIC SYNDROMES

One or more peripheral blood cytopenias with prominent maturation abnormalities in the BM (dysplasia)

MDS

- >50 years old
- 60% male
- Fatigue, weakness

PERIPHERAL SMEAR

- RBC- macro, dimorphism, basophilic stippling, NRBC, Howell-Jolly, Sideroblasts, Aniso, Poik, Retic
- WBC – Neutopenia, hypo granular, pseudo-Pelger-Huet, monocytosis
- PLT – thrombocytopenia, giant forms, hypogranular, micromegakaryocytes (dwarf megakaryocytes)

BONE MARROW

- RBC – megaloblastic, nuclear fragmentation, defective hemoglobinization, vacuolization, ringed sideroblasts
- WBC – Abnormal granules in promyelocytes, no secondary granules, nuclear abnormalities, ↓ myeloperoxidase, Auer rods in blasts
- Plt – Micromegakaryocytes, megakaryocytes with multiple separated nucleoli, hypogranulation or Ig abnormal granules

OTHER LABORATORY

- Iron – normal or ↑
- TIBC – normal or ↓
- Vit B12 and Folate – normal or ↑↑
- T lymphocytes ↓

FAB REFRACTORY ANEMIA

- RBC – macrocytic, ↓ retic, WBC & Plt normal
- BM – Hypercellular with erythroid hyperplasia, dyserythropoiesis, Blasts <5%

FAB REFACTORY ANEMIA WITH RINGED SIDEROBLASTS

- Similar to RA but >15 % of BM nucleated cells ringed sideroblasts
- < 5% Blasts, Hypercellular

FAB REFACTORY ANEMIA WITH EXCESS BLASTS

- Cytopenia in two cell lines
- <5% blasts in peripheral blood
- BM \leq 20% blasts

FAB REFACTORY ANEMIA WITH EXCESS BLASTS IN TRANSFORMATION

- \geq 5% Blasts in peripheral blood
- BM 20-30 % Blasts with or without auer rods, Variable sideroblasts



WHO CLASSIFICATION

- Refractory Anemia
- Refractory Anemia with Ringed Sideroblasts
- Refractory Cytopenia with Multilineage Dysplasia
- Refractory Anemia with Excess Blasts
- Myelodysplastic Syndrome with isolated del(5q)
- Myelodysplastic Syndrome, Unclassified



CHRONIC MYELOMONOCYtic LEUKEMIA

- < 5% blasts in peripheral blood, > 1 X 10⁹ monos
- BM ≤ 20% blasts, variable ringed sideroblasts, ≥monos



VARIANTS OF MDS

- Hypoplastic MDS
- MDS with Fibrosis
- Unclassified
- Therapy-related Myelodysplasia
- The 5q- Syndrome
- Childhood MDS



PROGNOSIS AND THERAPY

- <2 Yr Survival
- Leukemic Transformation
- Supportive Care – Transfusions, antibiotics
