

P1. CASE PRESENTATION: PROGNOSTIC FACTORS, CLASSIFICATION, AND RISK MODELS FOR OVERALL SURVIVAL PREDICTION IN MDS PATIENTS.

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The myelodysplastic (MDS) are a very heterogeneous group of myeloid disorders characterized by peripheral blood cytopenias and increased risk of transformation to acute myelogenous leukemia (AML).

Prognosis of MDS patients can be predicted by different risk models. Some are disease-related (FAB, WHO, IPSS, WPSS, MDACC) and some are patient-related (MDS-CI):

- FAB and WHO classifications do not account for cytogenetics
- IPSS is the best model for newly diagnosed patients, but can underestimate transfusion dependency and cytogenetics
- WPSS can be used at any time during the disease course, but can underestimate cytogenetics
- MDACC is dynamic and can be applied to patients with prior therapy, secondary MDS, CMML
- MDS-CI assesses the additional risk of comorbidities

We present the case of a male patient of 65 years old, diagnosed in Nov 2009 with MDS–RCMD with RS (WHO 2008); IPSS Int-1; WPSS Int; MDACC Low risk; MDS-CI Low risk.

The patient was transfusion – dependent, with 3-4 units /month.

After 12 months (Nov 2010): he was symptomatic with macrocytic anaemia (Hb 7.9 g/dL) and ferritin levels: 1121. No LIC or cardiac T2 available; No test for HFE gene mutation; No clinical signs of haemochromatosis. The treatment was Erythropoiesis-stimulating agent and Red cell transfusion + iron chelation.

September 2012: The bone marrow aspirate shows erythroblastopenia, multilineage dysplasia and 25% ring sideroblasts; Chest CT scan shows no modifications. He starts Cyclosporine 5 mg/kg – 8 mg/kg with no hematological response.

January – May 2013: Supportive care with transfusions: 6 units/month and iron chelation
Progressive heart failure appear - The patients is now MDS-CI Int - High risk. ECOG 3.

May 2013: The bone marrow aspirate shows erythroblastopenia, multilineage dysplasia and 20% ring sideroblasts; WPSS score: Intermediate
The Immunosuppression and transfusion – dependence influence the prognosis. In June 2013 the patient diagnosis is stafilococcal endocarditis and he dies, with no sign of leukemic progression.

It has become apparent that the natural history of patients with lower risk disease is very heterogeneous. Presence of comorbidity had a significant independent impact on survival and a prognostic score could be developed that assesses the additional risk of comorbidities.

IPSS is now replaced by a new revised score (IPSS-R) and by the incorporation of new molecular markers recently described.