

Myelodysplasia and its Frontiers

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Myelodysplastic syndromes (MDS) are considered clonal proliferations with a starting point in a multipotential haematopoietic mother cell (CD34+, CD117+).¹ They are typically defined as peripheral cytopenias evolving with a hypercellular bone marrow and with dysplasia signals in one or more series.¹ For a long time that the French American British Cooperation Group has proposed a classification based in semiquantitative morphologic criteria, obtained from observing peripheral blood and bone marrow swabs, stained with Romanowsky type colorations and iron specific.¹ Thus, 5 subtypes can be differentiated: Refractory Anemia (<5% blasts (bl); >15% sideroblasts in ring, in the bone marrow), sideroblastic anemia (5% bl; >15% sideroblasts in ring in the bone marrow). Refractory anemia with blasts excess (bl>5% and <30% in bone marrow), Refractory Anemia in transformation (bl>20% and 30% in bone marrow; or bl 5% in peripheral blood; or blasts with Auer bodies in peripheral blood and bone marrow) and Chronic Myelomonocytic Leukemia (bl>5% <30% in the bone marrow and monocytes > 1,000/mm³).²

This classification, universally accepted, starts now to be insufficient. There are ever more cases referred in the literature as “unclassifiable”, according to the above mentioned criteria.

We think it is important to mention: the 5q- syndrome, evolving typically with a normal or a high number of platelets and invariably with the deletion of the chromosome 5 long arm; as MDS with hypocellular bone marrows, particularly frequent after a bone marrow transplant; as MDS with myelofibrosis and myelosclerosis, and the overlapping syndromes

between MDS and myeloproliferative syndrome (MPS) where there are clinical and laboratorial findings related with both disorders. In this last group, Chronic Myelomonocytic Syndrome is included as, in spite of defined in FAB classification as MDS is worthy of being noted also in other systems due to their specificities as being a MPS. In fact, this is the disorder “by excellence” that can be found between two big pathologies groups.^{1,2}

Each one these unclassified cases has specific clinical laboratorial data, evolving also with very variable prognosis.^{1,2,3}

Most MDS patients was until sometime ago classified only on a clinical basis and in classic supplementary exams. It is already demonstrated, that in some cases, that is not possible, reason why only more sophisticated studies, probably with a clinical, morphologic and cytogenetic correlation can lead to a set up of new systematization criteria and eventually more effective specific therapies. ■

References

1. Kampmeier P, Anastasi J, Valdmeier JW. Issues in the Pathology of the myelodysplastic Syndromes. In: Myelodysplastic Syndromes. Hematology Oncology Clinics of North America. Koeffler PH. Philadelphia: Saunders; 1992:501-522.
2. Galton DAG. The Myelodysplastic Syndromes. In: Postgraduate Haematology. Hoffbrand AV editors. 3th ed. Oxford: Heinemann Professional Publishing Ltd. 1989:463-473
3. Gatterman AC, Gernig HA, Ferigs G et al. Myelodysplastic syndromes. Analyses of prognostic factors in 235 patients and profiles for an improved scoring system. Leukemia. 1992; 6:52-59