C9. REFRACTORY ANEMIA WITH RINGED SIDEROBLASTS AND THROMBOCYTOSIS WITHOUT JAK2 V617F MUTATION- REPORT OF THREE CASES

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Abstract: In the WHO classification, there is a provisional entity called - Myelodysplastic / Myeloproliferative Neoplasm, Unclassifiable (MDS/MPN,U). Refractory anemia with ringed sideroblasts associated with marked thrombocytosis (RARS-T) was included in this category. Recently published studies report a small percentage of patients with RARS-T. Sixty percentages of these have JAK2 V617F mutation, which can suggest the coexistence of two pathological conditions (MDS and MPN). In this paper, we analyzed three patients diagnosed with RARS-T in Hematology Department, Fundeni Clinical Institute, during the period 2005-2011. The patients were investigated with cytogenetic exam and molecular biology. In these three cases were identified morphological features of multilineage dysplasia (two lineage dysplasia in two cases and three lineage dysplasia in one case). In two cases thrombocytosis was under 1000 x 10^{3}/μl and clinical evolution was similar to the myelodysplastic syndrome (transfusion dependent anemia with response to administration of erythropoietin). In the third case, the platelets were over 1000x10^{3}/μl and with respons to the treatment with Hydrea, which improved anemia. JAK2 V617F mutation was not identified in any case.
RARS-T remains a provisional entity and requires a complex investigation of patients for the correct diagnosis of these patients. Therapeutic options should be personalized to each case in part because there is not yet a standardized treatment of these patients.