treatment because of lack of medicine in Unified Health System (SUS) demonstrates a treatment failure. **Conclusion:** Monitoring the progress of MDS in the patient was able to detect CMML at the beginning and already start the treatment.

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NFKB, TRAF6, AND MYDDOSOME GENE EXPRESSION IN MYELODYSPLASTIC NEOPLASM (SMD) AND ACUTE MYELOID LEUKEMIA MYELODYSPLASIA RELATED (AML-MR)

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Myelodysplastic neoplasms (MDS) are myeloid neoplasms, with bone marrow dysplasia, cytopenias, and increased risk of Acute Myeloid Leukemia (AML). Genotoxic agents and chronic inflammation can promote mutations, with clonal hematopoiesis and genetic instability. Chronic autoimmune and inflammatory disorders increase the risk of MDS. Several molecular and genetic mechanisms are involved in chronic inflammatory signaling of MDS, especially the activation of Toll-Like Receptors (TLRs), whose main intracellular adapter protein, MYD88, combines with IRAK kinases family (IRAK1, IRAK2, and IRAK4) in a complex called Myddosome, promoting TRAF6 phosphorylation and activation of NFKB and MAPK pathways and cytokine secretion. This study evaluated TRAF6, NFKB, and myddosome genes (MYD88, IRAK1, IRAK2, and IRAK4) expression in patients with MDS and AML Myelodysplasia Related (AML-MR), and its association with clinical variables and karyotype. Eighty-two patients, (76 with MDS and 6 with AML-MR) and 10 healthy controls were included. The cytogenetic study was carried out using G-Band and real-time PCR (RT-qPCR) methodology to analyze gene expression. IRAK4 and NFKB expression were higher in MDS with increased blasts (MDS-IB) compared to MDS with low blasts and ring sideroblasts ($p < 0.001$ and $p = 0.026$, respectively). MYD88 expression was lower in AML-MR patients compared to controls ($p = 0.041$), lower in patients with hemoglobin levels below 8 g/dL compared with hemoglobin levels above 10 g/dL ($p = 0.036$), lower in patients with bone marrow blast percentage > 10% compared to ≤ 10% ($p = 0.027$) and lower in patients with complex karyotype compared to normal karyotype ($p = 0.005$). IRAK1 expression was lower in patients with bone marrow blast percentage > 10% compared to ≤ 10% ($p < 0.001$) and lower in patients with abnormal karyotype compared to normal karyotype ($p = 0.031$). IRAK2 expression was lower in patients with bone marrow blast percentage > 10% compared to IPSS blasts categories (≤ 2%, > 2-< 5%, 5-10%) ($p = 0.007$, $p = 0.043$, and $p = 0.012$, respectively). Lower expression of MYD88, IRAK1, and IRAK2 was found in patients with adverse prognostic clinical features. Studies describe hyperactivity of TLRs pathways and their downstream molecules mainly in the initial phases of disease or low-risk MDS, while in high-risk diseases and advanced phases, reduced immune responses contribute to clonal evolution and progression to AML. Higher expression of IRAK4 and NFKB were found in patients with adverse prognostic clinical features, despite the downregulation of myddosome pathway, suggesting involvement of alternative pathways. In thiscohort, anomalous expression of NFKB and innate immune genes were observed in MDS and AML-MR patients, suggesting downregulation of myddosome pathway in the presence of adverse prognostic features, as well as hyperexpression of NFKB and IRAK4, both critical in MDS pathogenesis.

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DIAGNÓSTICO PRESUNTIVO DE NEOPLASIA MIELODISPLÁSICA ASSOCIADA A SÍNDROME VEXAS EM UM HOSPITAL PÚBLICO

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Introdução: Neoplasias Mielodisplásicas (SMD) correspondem a uma entidade hematológica responsável por alterações morfológicas em células medulares, resultando em citopenias, além de possuir risco de evolução para Leucemia Mieloide Aguda. Exposição quimioterápica prévia, exposição a substâncias mielotóxicas e doenças autoimunes são fatores de risco estabelecidos. Dentre as causas autoimunes, a Síndrome VEXAS (vacuolização, enzima ativadora de ubiquitina E1, ligação ao X, autoinflamação, síndrome somática) deve ser considerada. **Relato de caso:** Paciente masculino, 73 anos, previamente hipertenso. Admitido em nosso serviço devido a quadro de edema articular, inicialmente sem artralgia associada, perda ponderal de 10 quilos e pancitopenia com 5 meses de evolução. Nos exames de imagem, observado esplenomegalia e opacidades pulmonares em vidro fosco, podendo corresponder a pneumopatia de pequenas vias aéreas. Realizado aspirado de medula óssea, com mielograma hipercelular, exibindo displasia nas três linhagens, incluindo a presença de vacuolização de citoplasma nas células precursoras. A imunofenotipagem comprovou as alterações sugestivas de mielodisplasia. Exames laboratoriais com fator reumatóide e anti-Ro positivo em altos títulos. Aproximadamente 30 dias