anemia hemolítica autoimune é extremamente incomum. No entanto, marcadores de hemólise identificados nos exames laboratoriais juntamente com a exclusão de possíveis causas hereditárias podem corroborar com essa hipótese. Além disso, a melhora clínica observada com o uso de corticoterapia em dose imunossupressora, conforme preconizado na literatura, reforça essa associação. Este caso enfatiza a importância de relatar apresentações atípicas de doenças bem estabelecidas, especialmente quando há uma resposta positiva ao tratamento padrão. Além disso, este relato enfatiza a necessidade de considerar a AHAI ao diagnóstico mesmo na presença de resultados negativos no teste de Coombs direto.

https://doi.org/10.1016/j.htct.2024.09.139

BLOOD MARKERS RELATED TO INTRAVASCULAR HEMOLYSIS AND PAIN CRISES IN SICKLE CELL ANEMIA

LR Pereira, PPD Nascimento, CRB Domingos

Universidade Estadual Paulista Júlio de Mesquita Filho (UNESP), São José do Rio Preto, Brazil

Introduction: Sickle cell anemia (SCA) is a hereditary hematologic disorder resulting from the homozygous inheritance of hemoglobin S (Hb S). It is characterized by the polymerization of Hb S, which in turn causes an acute inflammatory state, chronic hemolytic anemia, vaso occlusive crises and other complex clinical manifestations. Intravascular hemolysis (IH) is considered a risk factor for severe clinical conditions in SCA due to the release of cell-free plasma Hb and its toxic effects. Also, one of the major clinical features of SCA is a acute pain crisis, which may lead to hospitalization due to the urgent need of intravenous treatment using opioids. This study aimed to evaluate the hemolytic profile as a risk factor for painful crises in SCA individuals. **Material and methods:** A total of 103 SCA patients in a steady state of the disease were included. Diagnosis confirmation occurred using classical analysis and the HPLC equipment Ultra 2® from Trinity Biotech. Hematological and hemolytic parameters were analyzed, and plasma cell-free Hb levels were quantified. Patients were stratified into two groups based on the frequency of painful crises in the past year (0-2 crises and ≥ 3 crises). **Results and discussion:** Cell-free plasma hemoglobin (Hb) levels were significantly higher in individuals experiencing three or more pain crises annually, while other markers showed no significant differences. Hematological parameters, though out of normal range, were consistent across subgroups, indicating chronic hemolytic anemia. Positive correlations were found between plasma Hb levels and reticulocyte percentage ($r = 0.769$; $p < 0.001$), as well as enzyme levels of AST ($r = 0.329$; $p = 0.001$) and LDH ($r = 0.772$; $p < 0.001$), with reticulocyte percentage having the strongest influence. The study underscores the role of cell-free plasma Hb levels as a direct marker of intravascular hemolysis (IH) and a potential prognostic parameter. Correlations with reticulocyte count and enzyme levels, particularly LDH and AST, further validate the importance of these markers in characterizing hemolytic profiles. Unconjugated bilirubin (UCB) did not correlate with plasma Hb levels but remains relevant in assessing extra-vascular hemolysis. **Conclusion:** Our study reinforces the critical role of plasma hemoglobin (Hb) as a marker of intra-vascular hemolysis (IH) in sickle cell anemia (SCA), particularly in relation to the frequency of painful crises. The significant correlation between plasma Hb levels and reticulocyte count, as well as with the enzymes LDH and AST, highlights the utility of these parameters in assessing hemolytic severity and guiding clinical management. Although unconjugated bilirubin (UCB) did not show a correlation with cell-free plasma Hb levels, it remains a valuable marker for extra-vascular hemolysis. These findings suggest that a comprehensive approach, incorporating both intra-vascular and extra-vascular hemolytic markers, is essential for understanding and managing the hemolytic profile in SCA. Further research, including genetic factors such as Hb F levels and βS haplotypes, could provide deeper insights into the variability of clinical outcomes and aid in the development of personalized treatment strategies. Monitoring and addressing hemolytic markers could offer significant benefits in reducing the frequency and severity of painful crises, ultimately improving the quality of life for SCA patients.

https://doi.org/10.1016/j.htct.2024.09.140

TERAPIA CLINÍCA DA COLESTASE INTRA HEPÁTICA AGUDA NA ANEMIA FALCIFORME: UM RELATO DE CASO

IT Melo, RC Cavalheiro, LMB Silva, FP Goncalves

Hospital Santa Marcelina, São Paulo, SP, Brasil

A colestase intra-hepática aguda (CIHA) representa uma complicação rara e grave da anemia falciforme (AF). Sua fisiopatologia consiste em falcização intra-sinusoidal, hipoxemia e isquemia dos hepatócitos com consequente edema dos hepatócitos e colestase intracanalicular, resultando em hepatomegalia e leucocitose, podendo desenvolver icterícia, disfunção renal, diátese hemorrágica e encefalopatia. O tratamento da CIHA consiste na transfusão de troca, tendo a eritrocitaferese maior segurança que a transfusão de troca manual devido à isovolemia no procedimento e resultados mais precisos de hematócrito e hemoglobina S (HbS). **Objetivos:** Relatar caso de tratamento clínico de colestase intra-hepática aguda em paciente com AF. **Métodos:** Trata-se de estudo descritivo, retrospectivo com aquisição dos dados em prontuário eletrônico, após consentimento do paciente. **Relato de caso:** Feminino, 42 anos, sem demais comorbidades, iniciou quadro de dor abdominal, náuseas, vômitos e icterícia há 1 semana, evoluindo com confusão mental, urina escurecida e hematomas extensos em membros superiores. À admissão apresentava Hb: 5,5, plaquetas 45.000, BT: 60 (BI: 33, BD: 27), INR: 2,7, acidose metabólica (pH:7,24/ HCO3:14,2), Cr: 1,7, UR:196, FA: 257, DHL: 1360, TGO: 67, TGP: 14. Tomografia de abdome evidenciava fígado de dimensões aumentadas, sem alterações obstrutiva e exames negativos para hepatites