citoplasmática, tornando as células sanguíneas sensíveis à lise pelo sistema complemento. Além de hemólise intravascular e consequente anemia, a HPN aumenta o risco de tromboses e insuficiência renal. A introdução do eculizumabe como tratamento da HPN de alta atividade melhorou o prognóstico e a sobrevida dos pacientes. O período gestacional, no entanto, devido à ativação do complemento que ocorre sobretudo no terceiro trimestre, cursa com alto risco de eventos tromboembólicos, morte materna, abortamento e parto prematuro. **Conclusão:** Apesar do prognóstico dos pacientes com HPN ter evoluído favoravelmente com o uso de bloqueadores do complemento, a gestação ainda é considerada de alto risco nessa população. O caso relatado ilustra que uma gestante com HPN, mesmo em uso regular de eculizumabe e tromboprofilaxia, não está livre de um evento desfavorável e ameaçador da vida.

https://doi.org/10.1016/j.htct.2022.09.092

ALENTUZUMAB AS THIRD LINE TREATMENT IN PURE RED SERIE APLASIA - A SUCCESSFUL CASE REPORT

MC Guedes, AVCDS Gasparine, TDS Baldanzi, FB Patricio, M Castro, MDC Gonçalves, R Marchesini, AP Azambuja

Hospital de Clínicas da Universidade Federal do Paraná (UFPR), Curitiba, PR, Brazil

Introduction: Pure red cell aplasia (PRCA) is a rare hematologic disorder characterized by reticulocytopenic, normocytic anemia with absence of red cell precursors in bone marrow, normal granulopoiesis, and megakaryopoiesis. The disease may be associated with viral infections, large granular lymphocytic leukemia, chronic lymphocytic leukemia, or thymoma, but the most common form is idiopathic. Refractory PRCA may respond to more profound immunosuppression with anti-CD52. **Purpose:** To describe a refractory PRCA patient treated with anti-CD52 humanized monoclonal antibody, alemtuzumab, with good response. **Case report:** A 52-year-old male patient with hemophilia A, previously treated with cryoprecipitate and currently using prophylactic Factor VIII, presents to healthcare with severe asthenia, no bleeding episodes during the period. Laboratory evaluation: Hb 3.4 g/dL; Ht 10.6% VCM 95; WBC 6.490 $\times 10^9$ /L; Platelets 196 $\times 10^9$ /L, intense reticulocytopenia (25 $\times 10^9$ /L). He had normal renal function, liver function, LDH, vitamin B12, folic acid, iron profile; serology with positive anti-HCV. Bone marrow evaluation showed 50-60% cellularity with intense hypoplasia of the erythroid series, granulocytic series with no changes in morphology or maturation pattern, and normal megakaryocytes with no signs of dysplasia. After investigation with immunoglobulin dosing, chest CT, flow cytometry for LGL, and serology for parvovirus, all normal, the diagnosis of constitutional APSV was confirmed. Treatment with cyclosporine (CSA) at a dose of 6mg/kg was initiated, with no reticulocyte response. In addition, he evolved with progressively increasing bilirubin levels (BT 7.4 BD 5.6) concomitant with a significant increase in Hepatitis C viral load (380,667 IU/mL Log: 5.58). Immunosuppression was discontinued, treatment with sofosbuvir and simeprevir was initiated, with good viral response. However, he required red blood cell transfusions every 2-3 weeks; new therapy was restarted with CSA and prednisone. The patient did not tolerate this treatment and developed severe thrombocytopenia and no improvement in hemoglobin levels. Considering the high risk of bleeding, CSA was discontinued. Platelet count normalized but he continued to require mensal RBC transfusion. At this point, second-line therapy with IV Alentuzumab, 3 mg dose followed by 6 weekly 15 mg doses (total 96 mg), coupled with infectious prophylaxis, has been proposed. Response occurred as early as the third infusion, as documented by a rapid increase in reticulocyte count. Within seven weeks the patient had a complete response (Hb 12.4 g/dL, Vg 35.3% and reticulocytes 170 $\times 10^9$/L). He had no toxicity-related intercurrences and remained with a complete response after six months of treatment. Unfortunately, a further progressive drop in hemoglobin after nine months from the end of the first treatment was observed. We opted to restart Alentuzumab at a dose of 15 mg/month, and follow with this maintenance dose. After the third application, the patient again showed a complete response (Hb 14.3 g/dL and VG 40.5%). Currently, the patient continues to use the medication every 45 days with a maintained hemoglobin level. **Conclusion:** Small case series cite Alentuzumab for refractory APSV as a treatment possibility with 63% overall response. We suggest use in refractory cases, followed by maintenance dosing to avoid relapse in long-term follow-up.

https://doi.org/10.1016/j.htct.2022.09.093

SEQUESTRO HEPÁTICO EM PACIENTE COM ANEMIA FALCIFORME: UM RELATO DE CASO

CVM Caxias, BSD Coelho, LG Siqueira

Universidade Estácio de Sá, Brasil

Objetivos: Apresentar um evento raro, em paciente com anemia falciforme, conhecido como: Sequestro Hepático. **Introdução:** A doença da anemia falciforme (AF) frequentemente leva a alterações hepáticas que podem ser classificadas entre agudas e crônicas. Alguns estudos sugerem que a disfunção hepática está mais frequentemente ligada à obstrução crônica de trato biliar ou a hepatite viral, isto é, causas não vasculares (CHARLOTTE, et al. 1985). No entanto, há também estudos indicando que são as alterações vasculares secundárias à doença falciforme desencadeiam lesões hepáticas e, em casos raros, o sequestro hepático (OMATA et al, 1986). **Materiais e métodos:** O relato se baseou em buscas de dados nos prontuários eletrônicos e laudos de exames laboratoriais e de imagem. **Relato de caso:** M.S.D., 40 anos, sexo feminino, dá entrada no serviço de emergência informando ser portadora de anemia falciforme, com astenia intensa, piora da icterícia, anúria e dor no quadrante superior direito do abdômen. Ao exame físico apresentava-se dispneica, taquicárdica, com edema bilateral em membros inferiores (4+/+4), ictérica (2+/+4), ritmo cardíaco regular em 2T, com bulhas normofonéticas, sem sopros, murmúrios vesiculares

presentes em ambos os hemitórax e creptos em bases somados à hepatomegalia importante, associada à dor à palpação do hipocôndrio direito. Nos exames admissionais foram constatadas anemia grave, sendo necessário realizar hemotransfusão com dois concentrados de hemácias. Além disso, foi percebido uma significativa leucocitose (28700/mm^3), DHL: 338,0 U nas Bilirrubinas (Bilirrubinas totais 3,81; Bilirrubinas diretas 3,7; Bilirrubinas indiretas 0,11 mg/dL) e na função renal (Ureia 33.8 mg/dL, Creatinina 1.4 mg/dL). Foi realizada ultrassonografia de abdome total que evidenciou hepatomegalia importante estendendo-se até o hipocôndrio esquerdo, baço não caracterizado, possivelmente relacionado a auto-esplenismo e derrame pleural à direita. Posteriormente, iniciou-se ácido fólico 5 mg/dia e furosemida para manejo do edema. **Discussão:** O sequestro hepático em pacientes com anemia falciforme é uma complicação rara e de difícil diagnóstico. Essencialmente clínico, é notadamente percebido dor em quadrante superior direito, associado a hepatomegalia no exame físico. Além disso, é percebido também a elevação dos níveis séricos da bilirrubina indireta, na desidrogenase lática, queda nas hemácias e hematócritos e elevação nas contagens de reticulócitos (BOTELHO, et al). O envolvimento da vesícula biliar é incomum, mas em casos em que existe constante secreção de bilirrubinas conjugadas aumentam as chances de desenvolvimento de colelitíase em pacientes homozigotos para a anemia falciforme e, devido a isso, faz-se necessário uma avaliação com a cirurgia geral para realização da colecistectomia, a fim de evitar novas complicações (TRAINA, et al. 2007). **Conclusão:** Pacientes com anemia falciforme que cursam com sequestro hepático devem ser considerados de alto risco devido às repercussões sistêmicas. O diagnóstico envolve, principalmente, uma anamnese detalhada, um bom exame físico associado e à achados laboratoriais. Nesse contexto, o prognóstico possui singularidades, sendo necessário uma abordagem multiprofissional (TRAINA, et al. 2007).

https://doi.org/10.1016/j.htct.2022.09.094

CLINICAL PREDICTORS OF VASO-OCCLUSIVE PAIN HOSPITALIZATION IN PATIENTS WITH SICKLE CELL DISEASE (SCD)

BA Oliveira [a], ICG Moura [b], MC Ozahata [c], SOG Mateos [d], AR Belisário [e], J Duarte [f], EC Sabino [a], S Kelly [g], CL Dinardo [h]

[a] *Instituto de Medicina Tropical, Faculdade de Medicina, Universidade de São Paulo (FMUSP), São Paulo, SP, Brazil*
[b] *Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, MG, Brazil*
[c] *Universidade de São Paulo (USP), São Paulo, SP, Brazil*
[d] *Universidade Municipal de São Caetano do Sul (USCS), São Caetano do Sul, SP, Brazil*
[e] *Fundação Centro de Hematologia e Hemoterapia do Estado de Minas Gerais (Hemominas), Belo Horizonte, MG, Brazil*
[f] *Fundação de Hematologia e Hemoterapia de Pernambuco (Hemope), Recife, PE, Brazil*
[g] *Vitalant Research Institute, San Francisco, United States*
[h] *Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo (HCFMUSP), São Paulo, SP, Brazil*

Introduction: Vaso-occlusive painful episodes (VOE) are the most common clinical manifestation of sickle cell disease (SCD) and the primary reason for hospitalizations and emergency visits. **Objectives:** This project aims to identify the clinical predictors of VOE hospitalization in adult participants of the Recipient Epidemiology and Donor Evaluation Study-III (REDS-III) Brazil SCD Cohort. **Material and methods:** The REDS-III Brazil SCD Cohort enrolled participants from 2013 to 2015 who were followed until 2018. Participants were randomly selected as eligible from public blood centers in 6 cities. Medical records were reviewed to abstract clinical data including the indications for all hospitalizations that occurred from one year prior to enrollment through the end of follow up. Adult cohort participants with 3+ hospitalizations per year due to VOE (cases) were compared to controls with less frequent VOE admissions to identify predictors of frequent VOE. **Results:** Of the 2793 participants enrolled in the cohort, 184 (6.7%) had 3+ VOE admissions per year (5.5% of 1330 children <18yr and 7.5% of 1453 adults). Compared to adults with <3 VOE admissions/ year, the adults with frequent VOE had higher white blood cell counts (11,566 vs. 98,659 cells/uL), platelet counts (389,500 vs 357,333 cells/uL) and lower hemoglobinF (4.4% vs 6.9%) and were more likely to have received red blood cell transfusions (80% vs. 41%) and hydroxyurea (60% vs. 49%) in the past year. Acute chest syndrome, bacteremia, and asthma were also more common in adult cases compared to controls (83% vs 61%, 16% vs 8% and 17% vs 8%, respectively). Independent predictors of 3+ VOE admissions/year in multivariable models included higher white blood cell (OR 1 [1.000-1.001], history of acute chest syndrome (OR 2.8 [1.581-5.202] and treatment with hydroxyurea (OR 1.8 [1.123-3.038]. **Discussion:** Frequent hospitalizations due to VOE are a marker of severe SCD used to identify patients for disease modifying therapy or even curative therapies such as bone marrow transplant. Understanding the significant predictors of frequent VOE in adults may identify other surrogate markers of severe disease that can be used to target therapies for high risk individuals. **Conclusion:** In this multi institutional cohort of SCD, white blood cell count and a history of acute chest syndrome were identified as significant independent predictors of frequent VOE while the association of hydroxyurea likely reflects treatment with this disease modifying therapy for frequent pain. Individuals with high white blood cell counts and acute chest syndrome should have close follow up to be monitored frequently and considered for early and aggressive treatment of pain as well as other disease modifying therapies.

https://doi.org/10.1016/j.htct.2022.09.095