81 anos, com relato de tuberculose pulmonar tratada com esquema RIPE (rifampicina, isoniazida, pirazinamida e etambutol) há cerca de 4 anos e diagnóstico prévio de hipertensão arterial sistêmica, hiperplasia prostática benigna e MFP com mutação JAK2, em tratamento com ruxolitinib. Apresenta esplenomegalia e trombocitose, embora com melhora clínico laboratorial. Foi internado devido à febre e sintomas respiratórios, com tomografia computadorizada (TC) de tórax evidenciando nódulos pulmonares sugestivos de metástase e nódulo em dorso sugestivo de abscesso. Foi iniciada antibioticoterapia empírica para infecção pulmonar bacteriana, com diminuição dos nódulos em nova TC de tórax pós-tratamento. A tomografia por emissão de pósitrons (PET-TC) realizada foi sugestivo de neoplasia em atividade, entretanto, a biópsia do nódulo pulmonar foi negativa. Exames laboratoriais evidenciaram anemia e trombocitose: hemoglobina 7,0 g/dL e plaquetas 550.000/mm^3, tendo sido realizada a transfusão de dois concentrados de hemácias. Consultas subsequentes evidenciaram manutenção da anemia, além de aumento da enzima lactato desidrogenase (429 U/L) e da eritropoietina (> 750 mIU/mL), além de Coombs direto positivo. Dado o diagnóstico de AHAI, iniciou-se o tratamento com corticoide e ácido fólico. **Discussão e conclusão:** Raros são os casos relatados na literatura de AHAI em pacientes com diagnóstico prévio de MFP. Este relato destaca a importância de documentação de tais casos para contribuir com a literatura médica e aprimorar a abordagem diagnóstica e o tratamento dessa condição complexa.

https://doi.org/10.1016/j.htct.2024.09.143

3.7 KB DELETION OF ALPHA-THALASSEMIA SIGNIFICANTLY ASSOCIATED FOR PRIAPISM IN SICKLE CELL ANEMIA PATIENTS FROM MANAUS-AM

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Objectives: Alpha thalassemia (AT) is a hereditary hemoglobinopathy caused by a partial or complete deletion of the synthesis of alpha globin chains. This deletion has been correlated as a prognostic marker which directly contributes to the phenotypic diversity demonstrated among sickle cell anemia patients (SCA). The aim of this study was to confirm the association of AT with clinical severity and decreased laboratory results in patients with SCA treated at HEMOAM. **Materials and methods:** A total of 156 male SCA patients with a mean age of 23+11 years old were molecularly investigated for AT for the presence of the 3.7 kb deletion. The molecular analysis was investigated by molecular technique of allele-specific PCR. Clinical and laboratory data were obtained from medical records and interviews with patients or legal guardians. **Results:** Twenty-six (16.7%) patients were heterozygous (-α/$\alpha\alpha$) and 11 (7.1%) were homozygous (α-/α-), totaling 23.8% of male patients with concomitant AT and ACS. Significant differences were demonstrated when analyses were performed between patients with wild type genotype for AT ($\alpha\alpha$/$\alpha\alpha$) and concomitant mutant's genotypes for AT (-α/$\alpha\alpha$ / -α/-α). Significantly decreased hematimetric indices for red blood cells ($p = 0.003$), mean corpuscular volume ($p = 0.023$) and mean corpuscular hemoglobin ($p < 0.1$), while elevated platelet count ($p = 0.032$) were observed in the mutant's genotypes, when compared to the normal wild type genotype. Interestingly, a higher prevalence of mutant genotypes (-α/$\alpha\alpha$ / -α/-α) was demonstrated in those with a higher presence of priapism, exhibiting a prevalence ratio of 4.48 (CI: 1.39-8.59 - $p = 0.018$). **Discussion:** We understand that the presence of AT may be related to lower frequencies of hemolysis among the group of patients with SCA, resulting in a higher concentration of hemoglobin and carriers of relatively higher hematocrit, favoring a higher blood viscosity, causing a higher frequency of blood blockage of the penile corpus cavernosum. **Conclusion:** Our results corroborate several studies that demonstrate AT as a phenotypic modulator for many of the severe comorbidities in patients with ACS, mainly in severe hemolytic crises, stroke and priapism, which is commonly correlated with the enormous clinical heterogeneity of sickle cell disease. The clinical course of patients with AF is highly variable and, although AT has been established as a modulator of AF, it still cannot explain all the clinical heterogeneity described among this group of patients. To try to clarify this genetic modulation, genotype-phenotype association studies, where candidate genes for single nucleotide polymorphisms are related to specific phenotypes, may help in this context. Genome-wide association studies of these SNPs may help confirm these observations and also describe other genetic modulators.

https://doi.org/10.1016/j.htct.2024.09.144

PÚRPURA TROMBOCITOPÊNICA TROMBÓTICA EM PACIENTE GESTANTE COM RESPOSTA APÓS CAPLACIZUMABE E IMUNOSSUPRESSÃO: UM CASO DESAFIADOR

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