remission and curing these patients. However, some patients might experience toxic effects as a consequence of variations in thiopurine metabolization, including TPMT and NUDT15 enzymes. In Brazil, there is a lack of studies that demonstrate the clinical association between drug toxicity in ALL treatment and genetic variants in genes related to thiopurine metabolism. The main objective of this study was to estimate the toxicity risk regarding polymorphisms of *TPMT* (rs1142345, rs1800460 e rs1800462) and *NUDT15* (rs116855232) in children with ALL. **Material and methods:** Pediatric patients diagnosed with ALL were included in the study. Recurrent gene fusions were identified by RT-qPCR and the copy number alterations were evaluated with MLPA. Genotyping was performed for *TPMT* and *NUDT15* genes using allele quantification assays with TaqMan probes. Hardy-Weinberg Equilibrium and odds ratio tests were conducted for statistical analysis of clinical and genetic data to assess the association between toxicity and genotype. **Results and conclusion:** A total of 154 individuals aged 0–18 years with ALL were analyzed, comprising 63 (40.9%) females and 91 (59.1%) males. Most patients (81.2%) had a common B-cell ALL subtype. Genotyping analysis showed heterozygous genotype frequencies of 7.8% for *TPMT* rs1142345, 2.6% for rs1800460 and 3.3% for rs1800462, while no variant genotype was found for *NUDT15* . Patients exhibited various manifestations of treatment toxicity, with febrile neutropenia (51.3%) being the most common. No significant association was found between genetic variants and toxicity symptoms. Considering that only patients with NM phenotype underwent thiopurine dosage adjustments during treatment, our study highlights the lack of clinical adherence to genotyping results for the thiopurine dosage adjustment in patients with the IM phenotype. This may impact their susceptibility to toxicity manifestations that might worsen their clinical condition. This study underscores the importance of pharmacogenetics in personalizing ALL treatment, offering potential to improve clinical outcomes by customizing thiopurine doses according to each patient's genetic profile. Supported by: CNPq, FAPERJ, Ministério da Saúde, Swiss Bridge Fundation.

https://doi.org/10.1016/j.htct.2024.09.1074

ACQUIRED HEMOPHILIA A IN A 3-YEAR-OLD PEDIATRIC PATIENT: A CASE REPORT OF RARE AND POTENTIALLY FATAL BLEEDING DISORDER

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Introduction: Acquired hemophilia A (AHA) is a rare severe autoimmune bleeding disorder with significant morbidity and mortality mainly occurring in older adults (average age 75). The condition is extremely rare in children. It is characterized by autoantibodies against coagulation factor VIII (FVIII), resulting in increased clearance and neutralization of FVIII leading to bleeding. Preventing and managing bleeding and inhibitor eradication are the mainstays of treatment. The outcome in pediatric patients seems more favorable than in adults because the inhibitors usually resolve more quickly and in a higher rate of patients. Yet we report a case of a child with AHA who had a lethal outcome 10 days post AHA diagnosis. **Case:** A 3-year-old female with history of fever, viral URTI and non-bloody diarrhea was diagnosed with parainfluenza infection. After two weeks, she developed progressive, total body edema with large ascites, and pleural effusions, mild acute kidney disease (creatinine 55 umol/L), hypoalbuminemia (albumin 22 g/L), elevated inflammatory markers (CRP 60.5 mg/L, ferritin 434.1 ug/L), and bicytopenia [normocytic anemia (Hgb 69g/L) with reduced reticulocyte count 36.2x109/L and reduced platelet count (17x109/L)]. She showed no findings of hemolysis, and peripheral smear showed only occasional schistocytes. Initially, she had a normal INR and fibrinogen but had a prolonged PTT at 83.3 sec which failed to correct with mixing study (she tested negative for lupus anticoagulant). After 4 days, the PTT prolonged further to 148.4 sec, and mixing study continued to show absence of correction. She started having some bruises, and a forearm hematoma where a peripheral IV was inserted. Further testing showed a FVIII level of < 1% whilst all other factors were within normal range. A high titer inhibitor (10 BU) was demonstrated by Bethesda assay. Extensive investigation was conducted to identify the underlying inflammatory condition, and differential diagnosis included atypical HUS, TTP (although ADAMTS-13 was normal), vasculitis and Castleman/TAFRO (given thrombocytopenia, anasarca, fever, renal dysfunction, elevated IL-6 at 11.6 pg/mL, as well as mild hepatomegaly and extensive, but small volume lymphadenopathy shown by abdominal CT). PCR testing for HHV-8 was negative. Bone marrow and lymph node biopsies were not pursued due to bleeding risk. She was started on rFVIIa 40 ug/kg (initially given prior to procedures, then increased to every 4 hours, then every 2 hours), and immunosuppressive therapy with intravenous methylprednisolone (30 mg/kg/day). PTT initially responded decreasing from 148 to 101 sec. However, on the third day after starting steroids, her PTT increased to 123 sec. On the fourth day of steroids, her left pupil was noted to be fixed and dilated. An urgent CT showed extensive bilateral subdural bleeds for which she underwent urgent decompressive craniotomy. Massive transfusion protocol was activated, and despite rFVIIa 90 ug/kg given every hour, tranexamic acid, platelet transfusions and FFP her subdural bleeding continued at which point further treatment was stopped and she passed. **Conclusion:** AHA is extremely rare in children but can be life-threatening. AHA should be suspected when a patient with no previous history of bleeding presents with bleeding, and an unexplained prolonged PTT. The awareness of such condition is essential for starting treatment. Despite treatment, poor outcome is still possible as seen in our case.

https://doi.org/10.1016/j.htct.2024.09.1075