condition, afebrile, and was discharged on the 2nd day of hospitalization, with clarithromycin (15 mg/kg/day). After G-CSF, ANC: 0.494×10^9/L (1st dose), 1.431×10^9/L (2nd dose). On outpatient follow-up, she had no long-term complications from Covid-19. Case Report 2: A man with chronic immune thrombocytopenia purpura (ITP) since 2008, autoimmune hemolytic anemia since 2013, evolved with AIN on May/2020, at 42 years old, with ANC lower than 0.5×10^9/L. On 6/1/2020, he had ANC 0.170×10^9/L. On 6/25/2020, he started flu-like symptoms, had ANC of 5.118×10^9/L, and tested positive for Sars-Cov-2 (RT-PCR). He kept high fever (102,2°F) and was hospitalized for 10 days without use of G-CSF. After discharge, on outpatient follow-up, he had no long-term complications from Covid-19, and presented ANC 0.338×10^9/L (Aug/2020). **Discussion:** At beginning of Covid-19 pandemic, severity infection in children was unknown. Today is known that most of them have milder clinical course, regardless of chronic diseases. In adults, in contrast, the inflammatory response tends to exacerbation, with more severe clinical conditions. Furthermore, many case reports of patients infected by SARS-CoV-2 with comorbidities literature are published. However, to date there are no reports on the impact of COVID-19 in AIN patients. Increased neutrophil counts during infectious episodes are common in AIN, which appears to be related to the benign course of most infections. We reported 2 cases of AIN patients diagnosed with Covid-19, both with favorable clinical outcomes despite heterogenic clinical course. On the first case, she presented few symptoms and ANC increased only after using G-CSF. On the second reported patient, there was a spontaneous increase of ANC and greater inflammatory response than the first case. It could suggest a correlation between inflammatory response to COVID-19 and ANC in cases of autoimmune neutropenia. **Conclusion:** In the reported cases, clinical course of disease and neutrophil count were different between adult and pediatric patients. It is not possible to state whether this difference is due to age group, individual response to infection or other variables. It is important to assess other cases of AIN infected by COVID 19 to better understand correlation between severity of infection and neutrophil count response.

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IS GFI-1 A TRIGGER TO DEVELOPMENT OF AUTOIMMUNE NEUTROPENIA?

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Objective: To describe a child carrying a *GFI1* -variant with autoimmune neutropenia. **Design/Method:** Single subject case report. **Results:** This is a case report of a 1 year-and-10-months-old female presenting severe chronic neutropenia ($0{,}03 \times 10^9$/L) with mild infections. The frequency of infections reduced as she got older. Physical examinations outside of infections were always normal and she had expected development. She did not have monocytosis or other hematologic disorders, and lymphocytes evaluation by flow cytometry analysis resulted normal. Viral diseases were negative, immunoglobulins dosage was normal for the age group, and screening for systemic autoimmune disease (sAID) was negative. Bone marrow aspirate analysis with karyotype resulted normal, without maturation arrest or dysplasia. Inborn errors of metabolism (IRM) and immunity (IRI) were ruled out. Anti-neutrophil (anti-HNA) autoantibody was detected on two occasions: at ages of 2-years-and-9-months-old and 5-years-and-3-months-old, presenting the same intensity and the same pan-reactivity to the FCγRIIIb glycoprotein. She has no siblings, but her father presented recurrent otitis until 6-year-old and hemogram with a persistent mild neutropenia documented since childhood. Ethnic benign neutropenia was ruled out. Both expressed Fyb on erythrocytes and were heterozygotes for single nucleotide polymorphism *rs2814778* in the *ACKR1*. She did not present remission of the disease until the age of 5 years and 7 months. A whole exome sequencing, showed a heterozygotic non-pathogenic *GFI-1* variant in exon 6 (c.929G>A:p.Arg310Gln). The variant, confirmed by Sanger sequencing, was also present in her father. He has no anti-neutrophil antibodies detected to date. **Discussion:** Most cases of pAIN are diagnosed between 3 to 8 months. Spontaneous remission in about 90% could be expected in 7 to 24 months of diagnosis or up to age of 4-5 years. In this reported case, the patient had severe neutropenia demonstrated since 1 year-and-10-month-old. Even 45 months after the beginning she remains neutropenic, carrying anti-HNA autoantibody with the same pan-reactivity features. The persistence of anti-HNA with no specificity may be associated to persistent autoimmune neutropenia (AIN) as well as a characteristic of secondary AIN, but screening for both sAID and IEI were negative. Due to the persistence of neutropenia, a whole exome sequencing was performed, presenting *GFI-1* variant inherited from her father, also neutropenic. The patient does not present features of *GFI-1* -variants, clinical course and the bone marrow were not compatible with SCN. In addition to regulating myelopoiesis, *GFI-1* also plays a role in the maturation of B and T lymphocytes, and in process of antigen-mediated lymphocyte activation. Although mature B-cells do not express *GFI-1*, this loss may be correlated with the increase of some IgG fractions, and some works have correlated *GFI-1* changes with autoimmune conditions and could be the cause of AIN reported. **Conclusion:** For patients with persistent pAIN, perfoming tests to detect a genetic involvement may be useful to understand the cause of the disease. In this case, the presence of *GFI-1* variant could be the trigger of AIN reported.

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AN UNUSUAL CASE OF NEUTROPENIA

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Objective: To describe a child with neutropenia and carrying inborn error of metabolism (IEM). **Results:** A 10 months-old boy, fist child of non-consanguineous parents, presented severe neutropenia (0.02×10^9/L) with fever, feeding problems, hypertonia and seizures. The frequency of fever and oscillating neutropenia is raised beyond use of antibiotics and granulocyte colony-stimulating factors. Some facial features present during physical examinations (hypertelorism, coarse face and skin dyspigmentation). He has failure to thrive and neuropsychomotor developmental delay. Viral diseases, organic acid in urine and mass spectrometry in tandem was normal. Abdominal ultrasound showed right hydronephrosis. Other image exams not detected alterations. The bone marrow aspirate analysis with karyotype resulted normal, with mild maturation arrest of granulocytic series. He did not have monocytosis or other hematologic disorders. Serum immunoglobulins were normal for the age group. Viral chronic diseases were negative, and she had no other autoimmune disorders. His mother had a first trimester spontaneous abortion, his father has no illnesses, and he has no siblings. A whole exome NGS sequencing showed compound heterozygous: one missense pathogenic *CLPB* variant (p. Arg408Gly) and other VUS c.1770+2T>A, splicing site. Hospitalizations is still frequent, and neutropenia did not remit to date. **Discussion:** This patient presents neutropenia and neurologic involvement, characteristics of caseinolytic peptidase B (CLPB) deficiency, which can range from severe to mild. In this reported case, a severe CLPB deficiency, death usually occurs at a few months of age due to neonatal neurologic involvement (hypotonia or hypertonia, respiratory insufficiency, swallowing problems, absence of voluntary movements, and epilepsy) and severe neutropenia associated with life-threatening infections. A multidisciplinary team is necessary, and no specific dietary or metabolic treatment is available. Hematopoietic stem cell transplant is not indicated in this case. Granulocyte-colony stimulating factor to increase neutrophil counts to reduce the frequency of infections should be used. Neutropenia associated with CLPB disease is present from birth and may be chronic or intermittent with absolute neutrophil counts ranging from severe (< 0.5 per mm^3) to mild (< 1.5 per mm^3). Elevated urinary excretion of 3-methylglutaconic acid (3-MGA) (typically 2x-10x higher than the reference range) has been observed in most of affected individuals to date. **Conclusion:** This case may be useful to understand the neutropenia in inborn error of metabolism and clinical signs and symptoms of the disease. The molecular evaluation is fundamental to elucidate diagnostic of neutropenia in newborns.

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LINFOMA DE HODGKIN

O PAPEL DA IMUNOTERAPIA NO LINFOMA DE HODGKIN

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O linfoma de Hodgkin clássico é derivado de uma complexa e imunomediada transformação maligna de linfócitos B originando células neoplásicas conhecidas como Reed-Sternberg (RS). Este tipo de célula sofre ativação dos fatores de transcrição NFkB promovendo o desenvolvimento e sobrevivência celular. Devido à recombinação e hipermutação genética das células RS, observa-se no cromossomo 9p24.1 a superexpressão dos ligantes de morte celular programada PDL-1 e PDL-2. Esses ligantes limitam as respostas imunológicas mediadas por células T ao desencadearem sua exaustão e perda progressiva de função. Dessa forma, as células tumorais que expressam ligantes PD-1 em sua superfície evitam uma resposta imune antitumoral eficaz. Atualmente de 20 a 30% dos pacientes com linfoma de Hodgkin clássico são refratários ou sofrem recaídas após tratamento envolvendo quimioterapia e ou radioterapia. A incorporação de novas terapias, incluindo inibidores de check point e conjugados de anticorpo-droga, no cenário da linha de frente, abordagens sequenciais e intensidades de tratamento individualizadas adicionais podem atender a essas necessidades. Por isso, essa pesquisa tem como objetivo reforçar a importância da incorporação de anticorpos monoclonais no tratamento do Linfoma de Hodgkin. Foram pesquisados nas plataformas de dados virtuais Scielo, PubMed e Google Acadêmico os atulos da literatura nacional e estrangeira e selecionados quinze de mais relevância sobre o tema. A quimioterapia de alta dose (HDCT) seguida por um transplante autólogo de células-tronco (ASCT) é o tratamento padrão para a maioria dos pacientes que apresentam recidiva após a terapia inicial. Para pacientes que falham HDCT com ASCT, brentuximabe vedotin, bloqueio PD-1, transplante alogênico não mieloablativo ou participação em um ensaio clínico poder ser considerados. Conclui-se que o bloqueio de PD-1 com pembrolizumabe tem demonstrado altas taxas de resposta e sem eventos adversos graves. Observa-se taxa de resposta objetiva em torno de 65% para os pacientes refratários ou recidivados pós-transplante medula óssea autóloga ou naqueles inelegíveis ao transplante.

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TREATMENT OUTCOMES IN CLASSICAL HODGKIN LYMPHOMA (HL): 5-YEAR UPDATE REPORT FROM THE BRAZILIAN PROSPECTIVE REGISTRY

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