

condition, afebrile, and was discharged on the 2nd day of hospitalization, with clarithromycin (15 mg/kg/day). After G-CSF, ANC: $0.494 \times 10^9/L$ (1st dose), $1.431 \times 10^9/L$ (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 $0.5 \times 10^9/L$. On 6/1/2020, he had ANC $0.170 \times 10^9/L$. On 6/25/2020, he started flu-like symptoms, had ANC of $5.118 \times 10^9/L$, 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 $0.338 \times 10^9/L$ (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 FC γ RIIIb 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, performing 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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