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Pregled | Review

Long Lasting and Late Onset neutropenia with and without antibodies against neutrophils. A potential new pathological entity?

Dugotrajna neutropenija i neutropenija s kasnim početkom sa i bez antineutrofilnih protutijela: Potencijalno novi patološki entitet?

Carlo Dufour[1]✉, Andrea Beccaria[1], Francesca Fioredda[1]

[1] Hematology Unit. Department of Hemato Oncology – IRCCS G. Gaslini Children's Hospital, Genova, Italy

Late Onset autoimmune neutropenias (LOAIN) are forms of neutropenia associated to detectable serum (indirect) anti neutrophil antibodies that rise beyond the age of 3 years. Long Lasting autoimmune neutropenias (LLAIN)are described as pediatric forms that do not remit within the classical time span of 24-36 months. LO/LL AIN were recently showed to have a different infectious pattern and a more pronounced autoimmune signature compared with primary autoimmune neutropenias (pAIN) that classically appear in the first year and remit within the third year of life and are associated to indirect positive anti neutrophil antibodies.

When no anti neutrophil antibodies are detected in the serum the LO/LL neutropenias are defined as idiopathic neutropenias (IN).

In the present study we analyzed, a large cohort of LO/LL AIN and IN patients included in the Italian Neutropenia Registry (INR) by performing extensive clinical, hematological, immunological investigations, and genetic analysis aiming to see whether there were differences between LO/LL AIN and IN or if they shared clinical, hematological and immunological features signature and a genetic background.

Indeed no major differences were detected between the two cohorts. A tendency to develop autoimmune phenomena was observed overtime in more than one third of the whole cohort. Specific alterations of B cells and T regulatory cells were observed in the cohort and pathogenic variants related to genes implicated in immune-dysregulations were found in a minority of patients.

Overall these findings suggest that LO/LL AIN and IN may represent a similar entity in which reduction of specific B cell subset and of T regulatory profile may predispose to development of autoimmune phenomena that in some case rely on an immune-dysregulation genetic background.

Young patients in whom autoimmune neutropenia does not remit in classical timespan should enter specific immunological investigation and monitoring plans.

✉ **Adresa za dopisivanje:**
Carlo Dufour, Hematology Unit. Department of Hemato Oncology – IRCCS G. Gaslini Children's Hospital, Genova, Italy