

PW03-034 - How to classify autoinflammatory diseases?

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MEETING ABSTRACT

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PW03-034 – How to classify autoinflammatory diseases?

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Introduction

Definitions and classifications of autoinflammatory diseases have been multiple. Their succession highlights the advances in our understanding of the innate immune system, especially the role of interleukin 1 β and the inflammasome. However, these definitions and classifications face a number of structure and content issues.

Objectives

To propose a novel definition of autoinflammatory diseases and to challenge the global classification of inflammatory diseases.

Methods

We appeal to the desirable characteristics of classification systems (exhaustiveness, disjointness, naturalness, usefulness) and to a critical analysis of the notion of continuum.

Results

We propose a clinically-oriented definition: “autoinflammatory diseases are diseases with clinical signs of inflammation, associated with elevated acute phase reactants and due to a dysfunction in the innate immune system, genetically determined or triggered by an endogenous factor”.

It is hard to find natural properties able to underlie a useful classification of autoinflammatory diseases, and inflammatory diseases as a whole, into disjoint and exhaustive categories. The notion of continuum is therefore appealing. However, a single continuum from purely autoinflammatory to purely autoimmune diseases oversimplifies, and even distorts, reality. How to locate, for instance, the disease caused by a deletion in *PLCG2* (the gene encoding phospholipase C γ 2) that associates autoinflammatory symptoms to both common variable

immunodeficiency and autoimmune features? Here we have an overactivation of both the innate and the adaptive immune system, associated with a deficiency of the adaptive immune system.

More than one dimension is needed to properly represent the immunological dysfunctions underlying inflammatory diseases. Furthermore, a classification of inflammatory diseases should also make sense of the clinical, pathological and biological phenotypes.

Conclusion

To be adequate and useful, a definition of autoinflammatory diseases and a classification of inflammatory diseases must take the multiple facets of reality into account, including clinical features. This can be done within a continuum only if it is multidimensional.

Disclosure of interest

None declared.

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