The Glycosphingolipid Hydrolases in the Central Nervous System

Massimo Aureli · Maura Samarani · Nicoletta Loberto · Rosaria Bassi · Valentina Murdica · Simona Prioni · Alessandro Prinetti · Sandro Sonnino

Received: 15 October 2013 / Accepted: 8 November 2013 / Published online: 27 November 2013
© Springer Science+Business Media New York 2013

Abstract Glycosphingolipids are a large group of complex lipids particularly abundant in the outer layer of the neuronal plasma membranes. Qualitative and quantitative changes in glycosphingolipids have been reported along neuronal differentiation and aging. Their half-life is short in the nervous system and their membrane composition and content are the result of a complex network of metabolic pathways involving both the de novo synthesis in the Golgi apparatus and the lysosomal catabolism. In particular, most of the enzymes of glycosphingolipid biosynthesis and catabolism have been found also at the plasma membrane level. Their action could be responsible for the fine tuning of the plasma membrane glycosphingolipid composition allowing the formation of highly specialized membrane areas, such as the synapses and the axonal growth cones. While the correlation between the changes of GSL pattern and the modulation of the expression/activity of different glycosyltransferases during the neuronal differentiation has been widely discussed, the role of the glycohydrolytic enzymes in this process is still little explored. For this reason, in the present review, we focus on the main glycolipid catabolic enzymes β-hexosaminidases, sialidases, β-galactosidases, and β-glucocerebrosidases in the process of the neuronal differentiation.

Keywords Glycosphingolipids · Neuronal differentiation and aging · Glycohydrolases · GBA2 · Neu3

Introduction

Along the differentiation process, the neuronal cells undergo important morphological and functional changes that contribute to their highly specialization. In these processes, the plasma membrane plays a fundamental role; in fact, modifications in its lipid and protein composition are responsible of the plasma membrane (PM) geometry thus generating specific signal transduction processes.

In particular, the PM of neuronal cells is characterized by a high content of gangliosides, glycosphingolipids (GSL)