Twenty years since the discovery of fulminant type 1 diabetes

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In 2000, fulminant type 1 diabetes (FT1D), which is characterized by a relatively low glycated hemoglobin level at onset of diabetes despite the abrupt occurrence of marked hyperglycemia with ketoacidosis, was discovered [1]. At the time of the discovery of this phenotype of diabetes, it was proposed to be a subtype of type 1B diabetes, idiopathic type 1 diabetes, because of the absence of islet-associated autoantibodies reported in the original paper [1]. However, islet-associated autoantibodies were detected in approximately 5% of FT1D cases in a nationwide study conducted later [2]. Moreover, recent reports have shown that anti-programmed cell death (PD)-1/anti-PD-ligand 1 (PD-L1) antibody therapy for malignancies leads to the development of FT1D, suggesting the involvement of autoimmunity in at least some proportion of FT1D cases [3]. Importantly, although it has been reported that FT1D was not observed in the Caucasian population in the early days [4], PD-1/PD-L1 antibody therapy-related FT1D did occur in Caucasians [3]; this fact lead to the consensus that FT1D does exist in the Caucasian population as well. Considering the importance of this subtype of type 1 diabetes, we would like clinicians and researchers to recognize this subtype globally. The following review will provide recent advances in FT1D, especially focusing on genetics, pathophysiology, and pathology, during the 20 years since its discovery.

Compliance with ethical standards

Conflict of interest A.S. has received lecture fees from Astellas Pharm Inc., Eli Lilly Japan K.K., Terumo Corporation, and Sanofi K.K. A.S. has received research funding from Astellas Pharm Inc. and Mitsubishi Tanabe Pharma Corporation. A.S. has received scholarship grants from Astellas Pharm Inc., Daiichi Sankyo Co., Ltd. and Kyowa Kirin Co., Ltd., MSD K.K., Novo Nordisk Pharma Ltd., and Ono Pharmaceutical Co., Ltd. H.I. has no financial conflicts of interest.

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