Niemann-Pick type C (NPC) disease is an autosomal recessive neurodegenerative disorder associated with mutations in NPC1 and NPC2 genes and characterized by an accumulation of unesterified cholesterol and glycosphingolipids in lysosomes. Deficiency of the corresponding NPC intracellular cholesterol transporter 1 protein thus leads to an abnormal cellular lipid and cholesterol composition. The most widely used NPC1 mouse model is the NPC1/− mouse that is characterized to display a strong neuroinflammation as shown by astrocytosis and increased levels of CD45-positive immune cells in the cerebellum and hippocampus.

To further evaluate levels and morphology of CD45-positive immune cells in NPC1/− mice, neuronal and visceral tissues were evaluated in comparison to wild type (WT) littermates. To this end, 10 µm thick cryo sections of the liver of 4 weeks old NPC1/− mice and the brain of 8 weeks old NPC1/− mice were assessed for CD45 expression by immunofluorescent labeling. In a next step the immunoreactive area was quantified in both the liver and brain tissue. Additionally, the numerical density, object intensity and object size of CD45-positive objects were evaluated in the thalamus and substantia nigra. Quantitative image analysis was performed with Image Pro 10 (Media Cybernetics) software and measured automatically so results are operator-independent and fully reproducible.

Immunofluorescent labeling of the liver showed a prominent enlargement of CD45-positive immune cells. Evaluating the overall immunoreactive (IR) area revealed that the signal was significantly increased when compared to WT littermates (Figure 1).

Our data suggest that NPC1 deficiency causes a severe neuroinflammation in the thalamus and substantia nigra as well as a severe change of hepatic leukocyte representation in the NPC1/− mouse model that perfectly mimics the pathology of the human lysosomal storage disease Niemann-Pick C1. These pathological hallmarks of NPC1/− mice will be especially valuable as readout in efficacy studies for the development of new treatments against Niemann-Pick disease.

**REFERENCE**


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