Supplementary Figure 1

Micrographs showing wt-misSOD1 staining in cerebellum (A-B), staining with an antibody against raised against whole SOD1 (C-D) and the effect of preincubation of the primary antibody with the immunizing peptide used as immunogen (E-H):
Sections from cerebellum (A, case #16) stained with the aa131-153 SOD1 antibody which reveal wt-misSOD1 stained glial cells surrounding degenerated Purkinje cells, enlarged in (B).
Sections from the spinal cord of a non-neurological control (C, case #61) and a patient with the *C9orf72HRE* mutation (D, case #16) stained with an antibody that equally detects whole natively folded wtSOD1 and misfolded wtSOD1: This antibody stains whole SOD1 as background staining in both the control (C) and the ALS patient with *C9orf72HRE* mutation (D). Small granular staining of wt-misSOD1inclusion might sometimes be discerned in the patient with *C9orf72HRE* mutation (D).
Blocking experiment in four sections from spinal cord motor neurons from a patient with a *C9orf72HRE* mutation stained with the aa131-153 SOD1 antibody (E-H, case #16). Several small granular wt-misSOD1 inclusions were seen in the soma when the antibody was preincubated only with diluent (E). The granular inclusions were weakly detectable when the antibody was preincubated with a low concentration of the immunizing peptide (F) and were barely detectable when the antibody was preincubated with intermediate concentration of the immunizing peptide (G). No wt misSOD1 inclusions were detected when the antibody was preincubated with a high concentration of the immunizing peptide (H).
Scale bars A, E-H 100 µm, B, D 50 µm, C 30 µm.