Accumulation of prion protein in the peripheral nervous system in human prion diseases

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摘要

Abstract

After the finding that anti-prion antibodies stain sensory and sympathetic ganglia in variant Creutzfeldt-Jakob disease (vCJD), it was suggested that this localization supported the oral route of entry. However, prion accumulation subsequently also appeared in the peripheral nervous system (PNS) in sporadic cases. This study aims at evaluating the extent of prion protein accumulation in the PNS in all clinicopathologic subgroups of the disorder, with the exception of the familial and sporadic forms of fatal insomnia. Patients included 2 vCJD cases, 2 Gerstmann-Sträussler-Scheinker (GSS), 2 iatrogenic (iCJD), and 16 sporadic CJD (sCJD) cases. Gasserian (17) and spinal (9), celiac (2) and thoracic sympathetic (one) ganglia, spinal cord and medulla of one vCJD, 2 GSS, one iCJD, and 5 sCJD cases were examined. Immunostained sensory ganglia were seen in both vCJD, both iCJD, one GSS, and 10 sCJD cases; the celiac ganglion was positive in one of two sCJD cases, and the spinal dorsal horn and the medullary sensory nuclei were positive in one patient with vCJD, one with iCJD, and 3 with sCJD. Western blot demonstrated presence of PrP in the gasserian ganglion of one patient with sCJD. Accumulation of prion in ganglia (including autonomic) of the PNS, shared by all subgroups of spongiform encephalopathy, and in the dorsal horns and medullary sensory nuclei, shows that the sensory route is involved in the trafficking of this protein