modes of inheritance, age at onset, and ethnic origin, the clinical heterogeneity of the disease and its clinicopathological overlap with other hereditary distal myopathies or oculopharyngeal dystrophy with rimmed vacuoles and amyloid deposits [3] suggests that mutations in exons 16 and 17 of this gene should be also searched in families with these disorders.

References

1. Dalakas MC. Polymyositis, dermatomyositis and inclusion body myositis. N Engl J Med 1991;325:1487–1498
2. Sekul EA, Dalakas MC. Inclusion body myositis: new concepts. Semin Neurol 1993;13:256–263
3. Mikol J, Engel AG. Inclusion body myositis. In: Engel AG, Franzini-Armstrong C, eds. Myology. New York: McGraw Hill. 1994:1384–1398
   Carpenter S, Karpati G, Heller I, Eisen A. Inclusion body myositis: a distinct variety of idiopathic inflammatory myopathy. Neurology 1978;28:8–17
5. Massa R, Weller W, Karpati G, et al. Familial inclusion body myositis among Kurdish-Iranian Jews. Arch Neurol 1991;48: 519–522
6. Mendell JR, Sahenk Z, Paul L. Amyloid filaments in inclusion body myositis: novel findings provide insights into nature of filaments. Arch Neurol 1991;48:1229
7. Askanas V, Engel WK, Alvarez RB, Glenner GG. β-Amyloid protein immunoreactivity in muscle of patients with inclusion body myositis. Lancet 1992;339:560–561
8. Askanas V, Engel WK, Alvarez RB. β-Amyloid precursor epitopes in muscle fibers of inclusion body myositis. Ann Neurol 1993;34:551–560
9. Goate AM, Chartier-Harlin CM, Mullan M, et al. Segregation of missense mutation in the amyloid precursor protein gene with familial Alzheimer's disease. Nature 1991;349:704–706
10. Naruse S, Igarashi S, Aoki K, et al. Mis-sense mutation Val-Ile in exon 17 of amyloid precursor protein in Japanese familial Alzheimer's disease. Lancet 1991;337:978–979
11. Mullan M, Crawford F, Axelman K, et al. A pathogenic mutation for probable Alzheimer's disease in the APP gene at N-terminus of β-amyloid. Nature Genet 1992;1:345–347
    Levy E, Carman MD, Fernandez-Madrid IJ, et al. Mutation of the Alzheimer's disease amyloid gene in hereditary cerebral hemorrhage. Dutch type. Science 1990;248:1124–1126
13. Sivakumar K, Dalakas MC. Quadriceps-sparing myopathy: an autosomal recessive, early-onset, familial inclusion body myopathy in non Iranian-Jewish families. Neurology 1995;45(S):209 (Abstract)
14. Yoshikai SI, Sasaki H, Doh-ura K, et al. Genomic organization of the human amyloid beta-protein. Gene 1990;87:257–263
15. Gajdusek DC, Beyreuther K, Brown P, et al. Regulation and genetic control of brain amyloid. Brain Res Rev 1991;16:83–114
16. Selkoe DJ. Normal and abnormal biology of the β-amyloid precursor protein. Annu Rev Neurosci 1994;17:489–517
17. Pericak-Vance MA, Bebout JL, Gaskell PC, et al. Linkage studies in familial Alzheimer's disease: evidence for chromosome 19 linkage. Am J Hum Genet 1991;480:1034–1050
18. Schellenberg GD, Bird TD, Wijsman EM, et al. Genetic linkage evidence for a familial Alzheimer disease locus on chromosome 14. Science 1992;258:668–670
19. Sarkozi E, Askanas V, Johnson SA, et al. β-Amyloid precursor protein is increased in inclusion body myositis muscle. Neuroreport 1993;4:815–818

Creutzfeldt-Jakob Disease after Liver Transplantation

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We report a 57-year-old woman who died from Creutzfeldt-Jakob disease 2 years after a liver transplantation. The liver donor had no history of neurological disease. In one albumin donor, possible Creutzfeldt-Jakob disease developed 3 years later. The patient initially had cerebellar symptoms. Neuropathology included "Kuru-type" plaques and prion protein (PrP) deposits involving the cerebellum predominantly. The patient was homozygote valine at codon 129 of the PrP gene while the liver was homozygote methionine. This observation raises the possibility of transmission of Creutzfeldt-Jakob disease by the graft itself or the associated albumin transfusions and, on a wider extent, by nonneural tissue.

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Since the first report of a corneal transplant transmission of Creutzfeldt-Jakob disease (CJD), the threat of iatrogenic transmission of CJD has been emphasized repeatedly. Neurosurgery, stereotaxic electroencephalography (EEG), corneal transplantation, dura mater implantation, tympanic membrane grafting, and injection of growth hormone or gonadotropin hormone extracted from cadaveric pituitary glands have been described as possible routes for prion transmission [1–3]. More recently, blood transfusions also have been suspected [4]. We report a case of CJD occurring 2 years after a liver transplantation (LT), raising the possibility of a new route for iatrogenic CJD.

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