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Epidemiological features of progressive multifocal leukoencephalopathy (PML) in Japan

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In order to clarify the epidemiological status of PML in Japan, We first defined a consensus diagnostic critera for PML: Possible PML (progressive encephalopathy with characteristic clinical features excluding other etiologies), Probable PML (features of possible PML plus JC virus DNA in CSF), and Definite PML (histological confirmation of typical features and JC virus infection). Based on the criteria a nationwide epidemiological survey revealed 52 cases (definite 20, probable 18, possible 14, M:F = 31:21) during these 5 years. Patients' backgrounds included AIDS (40%), collagen diseases (13%) and hematological malignancies (25%). Six cases had no identifiable evidence of immunosuppression. There was an increase in number of AIDS-related PML patients coincident with AIDS epidemic in Japan. The prognosis of most PML patients was poor with 5.6 months of median survival. However, some cases appeared to prolong survival with interferon, Ara-C or anti-HIV therapy. There were also a few patients with spontaneous remission. Efforts to elucidate the pathomechanism and develop effective treatments should be greatly increased.