Idiopathic CD4+ T-Lymphocytopenia

In 1983, human immunodeficiency virus (HIV) was detected as the pathogen of acquired immunodeficiency syndrome (AIDS). Thereafter, diagnostic procedures for HIV infection, such as cultivation of HIV, determination of serum antibodies against HIV viral antigens, detection of HIV viral genomes and reverse transcriptase, have been developed. As a consequence, the worldwide prevalence and the tendency for the increase of HIV infections have been clarified. Interestingly however, several cases of AIDS-like symptoms without HIV infection have been reported (1-4); such cases have been termed idiopathic CD4+ T-lymphocytopenia (ICL).

This situation is quite similar to the occasion when diagnostic techniques of hepatitis B virus infection became available. While hepatitis B virus infections were diagnosed in many cases of post-transfusion hepatitis, cases of chronic hepatitis without detection of hepatitis B virus infection were found. These cases led to the detection of hepatitis C or G viruses. Therefore, cases of ICL could be a clue for the detection of unknown pathogens for immunodeficiency syndrome.

Although the reports of ICL have been relatively few, an additional case was reported in the current issue of the journal by Hayashi et al (5). This case was complicated by Bowen’s disease with infection of human papilloma virus. To date, the cases of ICL were complicated with varieties of opportunistic infections, such as infections with Mycobacterium avium, Pneumocystis carinii, and fungi. Therefore, it is suggested that cases with opportunistic infections should be checked for possible complications of CD4+ T-lymphocytopenia.

Pathogenesis is one of the most serious questions regarding ICL. ICL does not always occur more frequently in populations with high risk factors for HIV infection. In addition, the decrease in CD4+ T-lymphocytes is not progressive and serum immunoglobulin concentrations are not elevated, compared with that of HIV infected AIDS, suggesting that the pathogenesis of ICL is different from AIDS with HIV infection.

The pathogenesis may not be a single factor, and various pathogeneses could be related to ICL (6, 7). Conventional viral infections also induce, at least temporarily, a decrease in CD4+ T-lymphocytes. And non-infectious diseases, such as common variable immunodeficiency syndrome or autoimmune diseases could be complicated with lymphocytopenia.

Interestingly however, there have been reports of human intracisternal A type retroviral particles found in cultures of peripheral mononuclear cells in ICL (8, 9). Serum antibodies against the particles were also detected in cases with ICL. These reports suggest the possibility that new types of retrovirus could be pathogens for ICL.

More reports of ICL should be accumulated, and international multi-center studies are necessary to clarify the pathogenesis and treatment of ICL in the near future.

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References