**CASE REPORT**

**Sarcoidosis and HTLV-1 infection**

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An asymptomatic, homosexual, white man was found to have an abnormal chest x-ray. A presumptive diagnosis of sarcoidosis was made, although pulmonary function tests and a transbronchial biopsy were normal. He remained asymptomatic for 10 years until he developed a progressive spastic paraparesis. At this point, antibodies to human T cell lymphotropic virus type 1 (HTLV-1) were identified in the serum and cerebrospinal fluid, and the diagnosis of HTLV-1 associated myelopathy was made, the history suggesting sexual exposure to HTLV-1 many years previously.

**DISCUSSION**

HTLV-1 was the first human retrovirus to be discovered and was initially reported as causing an aggressive T cell leukaemia/lymphoma.7 It was subsequently shown to be the primary cause of the progressive myelopathy previously called tropical spastic paraparesis, now known as HTLV-1 associated myelopathy (HAM/TSP).8 The virus is endemic in certain areas including Japan, the Caribbean, the Seychelles, and West Africa. Sporadic cases occur in Europe and the USA. Like HIV, transmission may be sexual, vertical, or via blood products.9 HTLV-1 is also associated with a spectrum of extraneural inflammatory disorders, attributed to the fact that the viral tax gene transactivates many human proinflammatory genes.
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development of both HAM/TSP and the extraneural immune normal.67 Unlike sarcoidosis, there are no granulomata, and a pronounced tropism for the lung, the most common lins, and oligoclonal bands in serum and CSF.4 The virus has and symptomatic infection is accompanied by systemic inflammation, with a raised erythrocyte sedimentation rate and C reactive protein, polyclonal increase in immunoglobulins, and oligoclonal bands in serum and CSF.7 The virus has a pronounced tropism for the lung, the most common extraneural association of infection being lymphocytic alveolitis, which is reported in up to 80% of individuals with HAM/TSP and 30–60% of asymptomatic carriers.3 The radiological abnormalities include appearances very similar to those of sarcoidosis.4 Pulmonary function tests may be normal.9 10 Unlike sarcoidosis, there are no granulomata, and HTLV-1 proviral DNA may be isolated from bronchoalveolar lavage fluid by polymerase chain reaction.4 Other associations of HTLV-1 include arthritis, hepatitis, skin rashes, sicca syndrome, and uveitis.3 A high viral load is related to the development of both HAM/TSP and the extraneural immune disorders.6

The association between HTLV-1 and sarcoidosis is complex. Clearly, given the spectrum of associated immune disorders, HTLV-1 infection may present with a syndrome very similar to sarcoid, and there are reports of cases in which sarcoidosis was diagnosed before the subsequent identification of HTLV-1 related aetiology.11 12 13 In addition to producing a clinical picture easily mistaken for sarcoid, a retroviral aetiology has been proposed in some cases of pathologically confirmed sarcoidosis, HTLV-1 proviral DNA having been isolated from sarcoid granulomata in the skin.12 13

“Human T cell lymphotropic virus type 1 associated myelopathy/tropical spastic paraparesis may progress rapidly in the presence of human immunodeficiency virus infection”

Although our patient presented with an illness clinically indistinguishable from sarcoidosis, the diagnosis was not confirmed histologically. Serum ACE was raised, but this is not specific for sarcoidosis, and although there are no published data on serum ACE in HAM/TSP, it is raised in patients chronically infected with HIV.14 Sarcoïdosis is believed to result from an exaggerated CD4+ T cell mediated immune response to a variety of antigens. In HIV related disease, the depletion of CD4+ T cells is thought to attenuate the formation of sarcoid granulomata, particularly when the CD4 count falls to below 200 x 10^6 cells/litre.15 The association between HIV infection and symptomatic sarcoidosis was considered unusual until the introduction of highly active antiretroviral therapy and recognition of the immune reconstitution syndrome. Our patient developed a myelopathy typical of HAM/TSP, with a history of sexual exposure to HTLV-1 many years previously. He was also HIV positive and although relatively immunocompetent, HAM/TSP may progress more rapidly in the presence of HIV coinfection.16 Our case illustrates the need for awareness of HTLV-1, the risk factors for seropositivity, and the spectrum of disorders associated with this proinflammatory retrovirus. With the benefit of this knowledge at the initial presentation, it would have been useful to have performed bronchoalveolar lavage with polymerase chain reaction for HTLV-1 proviral DNA, potentially enabling the diagnosis to be made earlier. Furthermore, although HIV was considered in the differential diagnosis of both clinical presentations from the outset, our patient’s racial and geographical origin meant that HTLV-1 was not considered until much later.

References


9 Osame M, Nakagawa M, Umehara F, et al. Recent studies on the epidemiology, clinical features and pathogenic mechanism of HTLV-1 associated myelopathy (HAM/TSP) and other diseases associated to HTLV-1. J Neurol Neurosurg Psychiatry 1997; 58(suppl 1): S30–1.


