A VERY RARE CASE – HAIRY CELL LEUKEMIA IN PATIENTS WITH SARCOIDOSIS


Two previous reports of accompanying HCL and sarcoidosis were noted in a patient with 12 years’ diagnosis of sarcoidosis with newly occurring HCL on the follow-up [5] and another one with a concurrent diagnosis [6].

Previous reports supposed that T lymphocyte defects were responsible for B cell proliferation into both HCL and sarcoidosis. In sarcoidosis, T cells recognize antigens and take part in amplification of local cellular immune responses which is achieved by the expression of various cytokine mediators. It has been demonstrated that T helper 1 cells exist in organs influenced by sarcoidosis. These cells produce IL-2 and IFN-γ and induce a nonspecific inflammatory response and granuloma formation [7]. Recent studies have shown the role of these cytokines in the development of HCL. Moreover, hairy cell activation includes expression of the autoregulated IL-2 receptor (the CD25 surface antigen represents the α-chain) [8]. So, the change in the cellular microenvironment by the sarcoidosis-defective T cells may contribute to development of HCL.

Although the cause of oligoclonal T cell proliferation in HCL has not yet been explained, it is similar to sarcoidosis. Sarcoidosis patients have a wide variety of oligoclonal T cells, and this represents response to different epitopes [7]. This makes one think that some epitopes triggering the inflammatory changes in sarcoidosis may contribute to hairy cell activation as exogenous stimuli.

It may be considered that development of sarcoidosis could be a result caused by HCL although one of the referenced case studies describes a temporal pattern of the two diseases. Some antigens restricted to hairy cells, such as CD11c and CD103, are associated with activation of lymphoid and non-lymphoid cell types [9].

In our case, the diagnosis of HCL was further confirmed by immunohistochemical staining of bone marrow biopsy. Whether it is only a coincidence or there is a causal relationship between HCL and sarcoidosis, merits further investigation.

References