CORRESPONDENCE

Sézary Syndrome With Warm and Cold Erythrocyte Autoantibodies

To the Editor:

Recently there has been interest in mycosis fungoides and the Sézary syndrome as examples of thymus-derived (T cell) lymphoproliferative disorders. The clinical manifestations, pathology, immunology, and tumor cell ultrastructure of both diseases have been well documented in recent reviews. However, little comment has been made of associated humoral abnormalities. In our review of 65 papers covering 817 cases of mycosis fungoides and the Sézary syndrome, only one paper commented on humoral abnormalities, and this was restricted to the presence of antinuclear and antitissue antibodies.

We recently cared for a patient with the Sézary syndrome who developed both warm and cold erythrocyte autoantibodies. We are unaware of a prior report of this finding with Sézary syndrome or mycosis fungoides.

Case. This 68-yr-old Portuguese woman was treated with prednisone and topical creams followed by a 3-mo course of chlorambucil for pruritus and cutaneous erythema. She was referred after 12 mo because of progression of her disease.

On physical examination her skin showed a generalized erythema and induration with multiple ulcerated nodules. She had bilateral axillary lymphadenopathy and mild hepatosplenomegaly. Laboratory data was notable for a hematocrit of 39.7, WBC of 8500/cu mm with 51 atypical lymphocytes, and platelet count of 156,000/cu mm. Her direct Coombs' test was positive, and her antinuclear antibody test was weakly positive, and her antinuclear antibody test was positive at a titer of 1:128, and the serum contained a small amount of cryoglobulin. Despite the presence of erythrocyte autoantibodies the patient did not manifest hemolysis.

We treated with high-dose methotrexate with citrovorum rescue. However, she died 10 days later of a cardiac arrhythmia.

Discussion. Several recent studies established mycosis fungoides and the Sézary syndrome to be lymphocyte disorders of the thymus-dependent cell line. The T cell nature of these neoplastic cells was suggested by their ability to form rosettes with sheep erythrocytes and to undergo blastogenesis in the presence of phytohemagglutinin. Serum from patients with these disorders was also found to contain significant quantities of macrophage migration inhibitory factor, a T cell lymphokine. Evidence that the Sézary cell is not a B lymphocyte includes a lack of membrane immunoglobulin and absence of receptors for the Fc fragment of IgG and receptors for the third component of complement.

Perhaps owing to the overwhelming evidence that mycosis fungoides and the Sézary syndrome are T lymphocyte diseases, very little has been recorded concerning humoral abnormalities in these conditions. Although antibody production is a B cell function, there is evidence that its production by B cells is modulated by T lymphocytes. It therefore seems possible that autoimmune antibodies would be a common finding in these patients. Nevertheless, almost no mention of autoantibodies is made in 65 papers on mycosis fungoides and the Sézary syndrome. Hence this appears to be the first report of erythrocyte autoantibodies in a
patient with the Sézary syndrome or mycosis fungoides. In contrast, patients with non-Hodgkin lymphoma and chronic leukemia have an incidence of erythrocyte autoantibodies of $1\times 10^5$, $2\times 10^5$, and $5\times 10^5$, respectively. Although we cannot prove that our patient’s erythrocyte autoantibodies were due to the Sézary syndrome, it is noteworthy that the amount of antibody increased in relation to exacerbation of her disease. A prospective study of the Coombs’ test in patients with mycosis fungoides and the Sézary syndrome will be needed to clarify the incidence of erythrocyte autoantibodies in T cell lymphoproliferative disorders.

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REFERENCES

3. Bibliography available on request from the authors of this Letter

ANNOUNCEMENT

The Second International Conference on the Adjuvant Therapy of Cancer will be held in Tucson, Arizona, March 28-31, 1979 under the sponsorship of the Cancer Center Division of the University of Arizona. Dr. Sydney E. Salmon and Dr. Stephen E. Jones will serve as Conference Co-Chairmen. The deadline for submission of abstracts (prepared in the format of AACR/ASCO) is December 1, 1978. For further information, write: Ellen Gerrity, Conference Coordinator, Cancer Center Division, University of Arizona, Tucson, Ariz. 85724.

ERRATUM

In the article Interaction Between Hb Hasharon and $\alpha$-Thalassemia: An Approach to the Problem of the Number of Human $\alpha$ Loci, by Pich, Saglio, Camaschella, David, Vaso, Ricco, and Mazza, published in the February 1978 issue [Blood 51:339-346, 1978], of the four symbols at the bottom of Fig. 1 (p. 340) the one farthest right should have borne the legend "$\alpha$-thal. heterozygote."
Sezary syndrome with warm and cold erythrocyte autoantibodies [letter]

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