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NCAM (CD56)-positive malignant lymphoma [letter; comment]

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NCAM (CD56)-POSITIVE MALIGNANT LYMPHOMA

To the Editor:

In a recent issue of *Blood*, Kern et al¹ described eleven cases of NCAM (CD56)-positive malignant lymphoma characterized by a predilection for extranodal involvement (central nervous system, muscle, skin, gastrointestinal tract, nasopharynx) and an aggressive clinical course. We are most delighted to read this report because we have independently made an identical observation.² In an analysis of the clinicopathologic features of nine patients with hematolymphoid malignancies characterized by the expression of CD56 (NCAM), unusual sites of involvement were also noted in our cases, including the skin and mucosal sites such as the lungs, jejunum, and salivary gland. We have not included nasopharyngeal/nasal lymphomas in our study because these lymphomas appear distinctive on their own, with a very high frequency of CD56 expression.³ An update of our experience shows that 27 (64.3%) of the 42 cases expressing one or more T-cell markers are CD56-positive (unpublished observation, July, 1992).

The immunophenotype of the cases in our series is practically identical to that reported by Kern et al.¹ The commonest phenotype is CD2⁺ CD3⁻ CD56⁺ CD16⁻ CD57⁻, and can be interpreted to represent true natural killer cell lineage² or peripheral T-cell lymphoma with CD56 expression,¹ although we favor the former

interpretation because genotypic study in one of our cases showed germline configuration of the T-cell receptor gene.

In our series, we had the opportunity to examine Giemsa-stained cytologic preparations in seven cases. Although the tumor cells show a broad range of cytologic appearances, cytoplasmic azurophilic granules can be demonstrated in all of them.² This cytologic feature provides a further link of the CD56-positive lymphomas with large granular lymphocytes, which comprise true natural killer cells and a subset of T lymphocytes.

To conclude, Kern's group and ours have independently shown the significance of CD56 (NCAM) in identifying a clinicopathologically and immunophenotypically distinctive group of aggressive malignant lymphoma. Therefore, it is important to include CD56 in the panel of antibodies used for immunophenotyping lymphomas.

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RESPONSE

We thank Drs Wong, Chan, and Ng for their comments regarding our report "Neural Cell Adhesion Molecule-Positive Peripheral T-Cell Lymphoma: A Rare Variant With a Propensity for

Unusual Sites of Involvement,"¹ and note with interest the publication of their series of similar cases.² Although there are slight differences between their series and our own, which might reflect

population or geographic factors, many of the salient features in the two series are similar.

Because of the small size of our series (11 patients) we did not attempt to separate the CD56/NCAM-positive patients into subgroups. In contrast to the experience of Wong et al,³ nasopharyngeal lymphoma is uncommon in our practice (<2% of the cases in our lymphoma collection). However, it is of interest to note that two of our three patients with nasopharyngeal involvement have had prolonged complete response (>4 years) and constitute the only patients in our group with long disease-free survival; this suggests that the nasopharyngeal patients may indeed be a separate group.

We were able to examine Wright-Giemsa-stained touch preparations in six of our cases; although occasional cells with large, azurophilic granules were seen in all cases they were present in a majority of cells in only one (a patient with nasopharyngeal involvement).

Assignment of this group of lymphomas to a definite lineage is difficult. All our cases were diagnosed as peripheral T-cell lymphomas based on routine histopathologic and immunophenotypic criteria.⁴ We hesitate to reassign them to natural killer (NK) cell lineage in the absence of other NK cell markers (CD16, CD57) and

the functional demonstration of NK activity; in addition, 18% expressed CD3, more consistent with T-cell lineage. One of our CD56-positive patients was studied by immunogenotyping; as in the experience of Wong et al, this case demonstrated germline configuration of both T-cell receptor and immunoglobulin genes. Although this might be felt to suggest NK rather than T-cell lineage, peripheral T-cell lymphomas lacking both β and γ T-cell receptor gene rearrangements have been described.⁵ Assigning lineage to this group of lymphomas will be an interesting intellectual challenge requiring further investigation; it is possible that they may be heterogeneous, with some cases demonstrating T-cell characteristics and others more closely resembling NK cells. At present it appears more important to recognize their potential for unusual biologic and clinical behavior, demonstrated in the series of Wong et al as well as our own.

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