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Peripheral T-cell lymphoma: prognostic factors

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The peripheral T-cell and NK/T-cell lymphomas are an uncommon and heterogeneous group of neoplasms with a generally poor clinical outcome. Studies of prognostic factors have largely been limited to comparative analysis by histologic type, and application of the International Prognostic Index (IPI) to heterogeneous groupings of these disorders.¹ In general, patients with anaplastic histology have had a better overall survival than those with non-anaplastic histology, and the IPI has been predictive of survival.^{2,3} Some studies have also found the IPI to be predictive in specific disease entities such as ALK-positive anaplastic large cell lymphoma^{4,5}, peripheral T-cell lymphoma unspecified^{4,6,7}, and nasal NK/T-cell lymphoma.^{8,9} Some studies have also found other clinical features to be predictive of survival including the presence of B symptoms, elevated β -2 microglobulin, or bulky disease, whereas other studies have found various pathological features such as the number of large transformed cells, proliferation rate, or EBV infection to be predictive.

To learn more about these various disease entities, a group of hematopathologists and clinicians recently undertook and completed a large

international study which included 1320 cases from 21 sites. One of the goals of this study was to evaluate various clinical and pathological features as potential prognostic factors within the various disease entities. Interestingly, the histologic diagnosis was highly predictive of overall survival ($p < 0.001$), with 5-year survivals ranging from 90% to only 7%. The IPI was also predictive of survival in all of the disease entities, with the exception of extranasal NK/T-cell lymphoma, and the distribution of the IPI scores generally correlated with survival.

In our study, we also evaluated the new prognostic model proposed by Gallamini *et al.*⁷ for PTCL-unspecified, but did not find it superior to the IPI. We also evaluated a new prognostic model proposed by Lee *et al.*⁹ for nasal NK/T-cell lymphoma and found it superior to the IPI. We also designed a new prognostic model for angioimmunoblastic lymphoma based on the three significant clinical predictors of survival in univariate analysis (age > 60 yrs, nonambulatory performance status, and B symptoms) and found it superior to the IPI.

We also found other clinical and pathological features in the various disease entities that had predictive value

Table 1. Survival in PTCL correlates with IPI scores.

	5-yr OS	IPI 0-1	IPI 3-5
Cutaneous ALCL	90%	86%	5%
ALCL, ALK ⁺	71%	51%	29%
Panniculitis-like	64%	42%	58%
ALCL, ALK ⁻	49%	41%	39%
Nasal NK/T	41%	41%	49%
PTCL-US	32%	28%	37%
Angioimmunoblastic	32%	14%	59%
Enteropathy-type	20%	25%	45%
ATLL	14%	19%	48%
Extranasal NK/T	9%	26%	60%
Hepatosplenic	7%	16%	84%

in univariate analysis, and multivariate analysis of these predictors along with the above prognostic models will be reported.

In conclusion, the IPI is useful for stratification of most patients with these disease entities for risk-adapted therapies. The superiority of the newly-proposed prognostic models needs to be confirmed in new prospective clinical trials. The evaluation of new clinical, pathological, and molecular genetic predictors in such trials is also needed in order to develop a more biologically-relevant prognostic model for each of the various disease entities.

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