

EPIDEMIOLOGY, CLINICAL FEATURES AND OUTCOMES OF PERIPHERAL T-CELL LYMPHOMA IN LATIN AMERICA

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Objective: To assess the distribution of Peripheral T-cell lymphomas (PTCL) across Latin America (LATAM) countries and report treatment outcomes. **Methods:** Patients aged ≥ 18 years with newly diagnosed PTCL from the retrospective registry of the Grupo de Estudio Latinoamericano de Linfoproliferativos (GELL, n=988, 1975-2023), T-cell Brazil Project (Brazilian TCP, ambispective registry 2015-2023, 593 cases) and the prospective International T cell Project (ITCP, n=529, 2006-2023). Survival data was available from the GELL and Brazilian TCP. Overall survival (OS) was from diagnosis to death from any cause, while progression-free survival (PFS) was diagnosis to relapse, progression, or death from any cause and it was applied Kaplan-Meier

method and Log-rank test. **Discussion:** Previous studies evaluating the distribution of PTCL subtypes across (LATAM) were limited in their representation of most countries in the region, however, now it was conducted an international pooled analysis and 11 countries of LATAM had enrolled. Besides that, a lack of standardized management for several subtypes and the absence of comprehensive lymphoma registries in LATAM suggests exploring real-world treatment patterns and clinical outcomes. **Results:** a total of 2110 pts from 11 LATAM countries were enrolled. Overall, the median age at dx was 54 years (18-95), 59% male, 67% had advanced stage disease and 29% had ECOG >1. The most frequently diagnosed PTCL subtypes was 39% PTCL not otherwise specified (NOS), 18% adult T-cell leukemia/lymphoma (ATL) and 16% extranodal NK/T cell lymphoma (ENKTL). Peru had a higher prevalence of ATL (39%) and ENKTL (43%) was frequently diagnosed in Central America. In contrast, ALK-negative (ALK-) anaplastic large T-cell lymphoma (ALCL) was the second in Brazil (18%), Chile (16%) and Argentina (9%); T-cell NOS was 28% in Chile and 27% in Argentina. First-line chemotherapy (CT) varied across subtypes and 77% received CT. Pts with ENKTL were frequently treated with asparaginase/platinum-based therapy (62%), while CHOP was used for ATL or PTCL NOS (46% for both). CT with CHOEP/EPOCH was used with ALK- ALCL (45%), ALK+ (47%), or AITL (48%). A median follow-up of 33 months, the 3-year OS and PFS were 40% and 30%, respectively. ALK+ ALCL had superior survival, with 77% OS and 73% of PFS. The ENKTL was 48% of OS and 45% PFS. Pts with ATL had the lowest survival rates (23% OS and 16% PFS). The use of asparaginase/platinum or CHOP-based therapy was associated with superior 3-year OS (61% and 52%, respectively; $p=0.011$) and PFS (57% and 49%, respectively; $p=0.017$) among pts with ENKTL. For ATL, the use of CHOEP/EPOCH was associated with improved 3-year OS (21%, $p=0.009$) and PFS (15%, $p=0.024$). **Conclusion:** To our knowledge, this is the largest pooled cohort of PTCL subtypes across LATAM. Our findings suggest a distinct distribution of PTCL subtypes, with a higher prevalence of ATL and ENKTL compared to the epidemiological patterns in Western countries. The relatively high percentage of PTCL-NOS suggests difficulties in providing specific lymphoma diagnoses in the region. The low survival rates for some subtypes indicate the need to develop novel therapies to improve patient outcomes. A larger prospective assessment of PTCL epidemiology and treatment outcomes is being planned to expand the ascertainment of cases, improve pathological classification of the different PTCL subtypes, and validate our results in the LATAM region.

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MIELOMA MÚLTIPLO ANAPLÁSICO: A IMPORTÂNCIA DA MORFOLOGIA NA ERA DA HEMATOLOGIA MOLECULAR

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Introdução: O mieloma múltiplo anaplásico (MMA) é uma entidade rara, com apresentação clínica agressiva e prognóstico adverso. O Mieloma Múltiplo (MM) pode apresentar a forma anaplásica desde o seu diagnóstico, ou se transformar durante seu curso. A suspeita diagnóstica se dá principalmente a partir da morfologia celular, tendo poucos casos descritos na literatura, o que dificulta seu entendimento e estudo, entretanto apresenta desfechos desfavoráveis à luz dos tratamentos atuais. **Objetivo:** Descrever um caso de mieloma múltiplo anaplásico acompanhado no Hospital Universitário Pedro Ernesto (HUPE). **Métodos:** Dados foram coletados de prontuário físico e eletrônico no HUPE. **Descrição do caso:** Paciente masculino, 62 anos, com primo diagnóstico de Mieloma Múltiplo IgA/Kappa em 2019, obtendo remissão parcial após 6 ciclos de CTD (Ciclofosfamida, Talidomida e Dexametasona) e consolidação com transplante autólogo de medula óssea (TAMO). Apresentou recaída em outubro de 2020, quando o mielograma demonstrava 12% de plasmocitose medular de morfologia habitual. Realizado resgate com 6 ciclos de VCD (Bortezomib, Ciclofosfamida e Dexametasona), obtendo resposta parcial. Permaneceu com doença estável até setembro de 2022, quando voltou a apresentar progressão de doença com anemia, hipercalcemia, lesões líticas difusas e aumento de proteína M. Foi iniciado protocolo VTD-PACE (adição de Platina, Ciclofosfamida, Doxorubicina e Etoposídeo), entretanto, o paciente desenvolveu cardiotoxicidade associada ao tratamento sendo continuado o tratamento com 4 ciclos de VRD (Bortezomib, Lenalidomia e Dexametasona) sem sucesso. O paciente evoluiu com progressão da doença, com piora da anemia e trombocitopenia, sintomas constitucionais, aumento de LDH (>3000 U/dL) e hipercalcemia sintomática. Novo mielograma com infiltração maciça de células plasmáticas de morfologia pleomórfica e anaplásica, de citoplasma basofílico, multinucleadas, com cromatina frouxa e presença de nucléolos evidentes. Imunofenotipagem confirmava células mielomatosas, com marcação positiva para CD138, CD38 e CD56, e negativa para CD117, CD19 e CD45. Sangue periférico sem presença de plasmócitos circulantes. Citogenética com hiperplóidia em 70% das metáfises analisadas. Optado por iniciar quimioterapia paliativa com ciclofosfamida oral e dexametasona e o paciente evoluiu a óbito. **Discussão:** O caso demonstra que, embora a forma anaplásica seja rara, ela deve ser considerada no diagnóstico diferencial e investigada, especialmente em pacientes com suspeita clínica de mieloma múltiplo com acometimento extramedular, citopenias e elevada carga tumoral com marcadores de turnover celular elevados. A variante anaplásica como apresentação primária ou progressão de doença está associada a um curso clínico agressivo, morfologia atípica, citogenética adversa, resistência à quimioterapia e baixa sobrevida em curto prazo. Algumas séries de casos escassas sugerem que a quimioterapia de indução agressiva, combinada com novos agentes, seguida de consolidação com TAMO deva ser considerada nesses pacientes. **Conclusão:** Este caso enfatiza o papel crucial da avaliação do mielograma de forma pormenorizada para o diagnóstico de neoplasias hematológicas, de modo a ajudar na inferência prognóstica e,