

massa em região malar, e foi iniciado tratamento quimioterápico com protocolo com dose reduzida de R-CHOP por dois meses. Após o tratamento, evoluiu com quadro de pneumonia e surgimento de novas massas ganglionares, sendo considerada refratária a essa linha de tratamento. Foi iniciado, então, tratamento com R-Clorambucil, evoluindo com piora clínica no segundo ciclo, após dois meses de tratamento, apresentando náuseas frequentes e manutenção das massas ganglionares. No terceiro ciclo, foi suspenso o clorambucil, finalizando o tratamento com 5 ciclos de rituximabe por dois meses, com resposta parcial, mantendo edema na face. Iniciou tratamento com acalabrutinibe por 6 meses, evoluindo com redução total das massas tumorais, porém interrompeu o uso por dificuldade de comprar a medicação. A paciente seguiu em acompanhamento com o serviço de hematologia, realizando uso irregular do inibidor seletivo de BTK (tirosina quinase de Bruton), sem novas massas tumorais, evoluindo para óbito por descompensação de DPOC 18 dias após última consulta. **Discussão:** O comprometimento extranodal do linfoma de células do manto afeta preferencialmente trato gastrointestinal e região de cabeça e pescoço, como o anel de Waldeyer. Embora o LCM seja uma condição rara, ele deve ser submetido a um diagnóstico diferencial quando afeta a área maxilofacial. Nesse contexto, exames de imagem, análises morfológicas, imuno-histoquímicas, citogenéticas e moleculares são fundamentais para chegar ao diagnóstico correto e levar ao tratamento adequado. **Conclusão:** O diagnóstico de LCM pode ser desafiador devido às suas semelhanças com outros tipos de LNH e apresentações em regiões atípicas podem tornar esse desafio ainda maior. Diante disso, torna-se essencial o relato de uma doença rara junto de uma manifestação ainda mais incomum.

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OUTCOMES OF PATIENTS WITH ADULT T-CELL LEUKEMIA/ LYMPHOMA (ATLL) IN A UNIVERSITY AND PUBLIC CANCER CENTER IN BRAZIL

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Introduction: Adult T-cell Leukemia/Lymphoma (ATLL) is a hematological malignancy caused by the Human T-cell lymphotropic virus type 1 (HTLV-1). ATLL has a broad clinical presentation, with a poor prognosis and limited therapeutic options. In 2012, the prevalence of HTLV-1 infection in Brazil was estimated at 83 to 222 cases per 100,000 blood donors, with 800,000 people expected to be living with the virus. Despite being an endemic country for the virus, the number of registered cases of ATLL is much lower than would be expected, indicating an underdiagnosed and poorly characterized disease in Brazil. **Methods:** This is a retrospective observational study conducted at HC-FMUSP. Our hospital has an outpatient clinic for the monitoring of asymptomatic HTLV-1 carriers, with 720 patients followed since 1994. We

describe the analysis of clinical, laboratory and epidemiological data collected from the medical records of patients diagnosed with ATLL from 1994 to 2022. **Results:** Overall, 55 patients were included and classified according to Shimoyama's classification groups. Median age at diagnosis was 52 years. Twenty nine patients (52%) were diagnosed with indolent (smoldering/chronic), 7 (13%) with lymphoma, and 18 (35%) with acute ATLL. Most patients diagnosed with indolent forms (22 out of 29) came from the cohort of asymptomatic HTLV-1 carriers. Most patients (51%) only discovered HTLV-1 infection at the time of ATLL diagnosis, this being particularly higher (76%) among patients diagnosed with an aggressive clinical form. For those patients followed at the outclinic, the median time between the diagnosis of HTLV-1 infection and the diagnosis of ATLL was 9 years (IQR 2-17). Fourteen patients with the smoldering/chronic disease evolved to an aggressive ATLL form, with a median time to progression of 2 years (IQR 0.8-3.8). These patients had a median overall survival of just 55 days after the diagnosis of the aggressive form. The median overall survival was 11 years for smoldering, 3.4 for chronic, 0.9 years for lymphomatous and 1.1 years for acute forms. A total of 35 patients received chemotherapy for aggressive disease, showing a low rate of response (20%), with CHOEP being the most commonly used protocol. Only 4 patients underwent allogeneic bone marrow transplantation, and one of them has the longest survival at the follow-up, at 7 years. **Discussion:** Having one of the largest HTLV-1 carrier populations worldwide, Brazil still needs better monitoring and characterization of ATLL. Even in a reference center where asymptomatic HTLV-1 infection is monitored, half of the cases were diagnosed already in an aggressive clinical form, with many patients only discovering HTLV-1 infection at the time of ATLL diagnosis. Patients who progressed from indolent forms of disease during follow-up had a very poor prognosis, similar to those initially diagnosed with aggressive forms, which is measured in months. Few patients are eligible for allogeneic bone marrow transplantation, the only curative therapy for the disease. **Conclusion:** Our findings highlight the underdiagnosis of HTLV-1 infection in Brazil and the lack of effective measures to prevent disease progression and treat aggressive disease forms. This emphasizes the importance of screening and preventive measures for viral transmission in our country.

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SÍNDROME DA CAUDA EQUINA COMO MANIFESTAÇÃO INICIAL DE LEUCEMIA DE BURKITT

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Objetivo: Apresentar um caso de HLH secundária à LB. Paciente masculino, 70 anos, com psoríase em tratamento com secuquinumab, internou por piora progressiva de parestesia