before the institution of up-front treatment for HHV-8-MCD. **Discussion:** HLH is a potentially lethal condition caused by macrophage hyperactivation following inflammatory, neoplastic or infectious triggers. Among the main causes, the HHV-8 virus highlights, particullarly in the context of immunosuppression. This virus is also involved in the pathogenesis of MCD and KS. There are, however, few reports of association between both disorders. Moreover, the association between MCD and TLS is also rare, since it is a non-neoplastic lymphoproliferative disease with low cellular turn-over. In view of the possible evolution to aggressive non-Hodgkin's lymphoma during the MCD's course, the TLS presence should prompt get to the search for concomitant DLBCL, which was not possible in this case due to the unfavorable premature evolution. **Conclusion:** This case showed that HLH and TLS, although rarely associated with MCD, can lead to serious organ dysfunctions with a potentially fatal course. Prompt recognition of both conditions is essential to establish early therapeutic measures that are crucial to ensure satisfactory clinical outcomes.

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TRANSFORMATION OF FOLLICULAR LYMPHOMA TO A BURKITT LYMPHOMA ASSOCIATED WITH HIV INFECTION AND PULMONARY TUBERCULOSIS

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Objective: To present a clinical case of follicular lymphoma (FL) transformed into Burkitt's lymphoma (BL) in a patient with acquired immunodeficiency, a rare case with little description in the literature. Methods: retrospective descriptive work of a clinical case carried out through data collection from HSPE medical records. **Results:** Woman, 62 years old, seeks emergency care due to generalized lymph node enlargement, fever, night sweats and weight loss. In the investigation, infection with the acquired immunodeficiency virus (HIV) and pulmonary tuberculosis was detected, and later there was the diagnosis of grade 3B FL, through a biopsy of the cervical lymph node. Positive immunohistochemistry for CD79A, CD20, BCL6 and KI-67 40%; negative for CD10, BCL2 and C-MYC. Treatment with RIPE and ART regimen was started and LF's outpatient follow-up was chosen. On CT scans, patient had stable lymph node enlargement with the largest measuring 2.1x1.6 cm in the left armpit. Seven months after the diagnosis, the patient had headache, arthralgia, abdominal pain and ecchymosis on the lower limbs. Tests showed hemoglobin (Hb) 12.5 g/dL, hematocrit (Ht) 36.6%, leukocytes (Lt) 17360/mm^3, atypical lymphocytes and platelets 15000/mm^3; lactic dehydrogenase (LDH) 13318 U/L, creatinine (Cr) 1.2 mg/dL, triglycerides 460 mg/dL, ferritin 2260 ng/mL, uric acid 17.8 mg/dL. Myelogram: 91% of small to moderately sized cells, high nucleus/cytoplasm ratio, loose chromatin, barely evident nucleoli and cytoplasm with intense basophilia without granulation and with microvacualization, suggestive of BL. Positive immunophenotyping for CD10, CD13, CD15, CD19, CD20, CD38, CD71, CD79a, CD81, cytoplasmic IgM, HLA-DR, Kappa. FISH with MYC-IGH gene rearrangement. CT with enlarged lymph node enlargement, the largest one in the left axillary 4.7 x 3.8cm. After 24 hours, the LDH was 22142 U/L, Cr 2.9 mg/dL, Hb:7.8 g/dL, Lt 57120/mm^3 with 45% blasts and platelets 25000/mm^3. Chemotherapy with R-CODOX-M was initiated, but the patient died on the 1st day of chemotherapy. Discussion: The most common histological transformation of LF occurs to a diffuse large B cell. Transformation into BL is uncommon. The subsequent molecular events that lead to transformation into a high-grade lymphoma are not fully understood, but a translocation leading to c-myc overexpression appears to be the trigger for transformation. BL may show c-myc-only abnormalities, association with bcl-2, or other karyotypic changes. In the presented case, we highlight an unusual clinical picture in a FL, with severe thrombocytopenia and increased LDH, without constitutional symptoms and/or visible enlargement of lymph nodes, which led to a spinal cord study and the diagnosis of transformation into BL. In one review with 4 cases, the median time to histologic transformation from follicular lymphoma to Burkitt's lymphoma was 6 to 13 months. In the case described, the patient initially had a FL, negative for BCL2 and C-MYC, which turned into a BL after 7 months of evolution, with the MYC-IGH rearrangement. Conclusion: As this is an uncommon evolution of FL, we emphasize the importance of suspecting transformation into high-grade B lymphomas in the face of unusual alterations in FL, such as severe cytopenias, constitutional symptoms, increased LDH and rapid progression of lymphadenopathy so that the treatment is instituted early.

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PEMBROLIZUMAB PLUS RADIOTHERAPY AS A RESCUE THERAPY FOR LANGERHANS CELL SARCOMA: A CASE REPORT

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Introduction: Langerhans Cell Sarcoma (LCS) is an aggressive and extremely rare form of malignant histiocytosis, with a dismal prognosis. **Case report:** A 62-year-old male with mild comorbidities (overweight, hypertension, type 2 DM and NAFLD) presented with chest and epigastric pain,

without constitutional or neurological symptoms or skin changes. His physical examination was unremarkable, and initial evaluation only revealed mild elevation of liver enzymes and C-reactive protein. Abdominal MRI revealed several hepatic nodular lesions associated with large peritoneal nodules in the hepatic hilum, adjacent to the pancreas and along the curvatures of the body and gastric antrum. Upper gastrointestinal (GI) endoscopy showed a large subepithelial lesion with ulcerations in the gastric antrum. PET-CT pointed to hypermetabolic cervical (measuring up to 19 mm, SUVmax 29.8) and abdominal lymph nodes (measuring up to 53 x 32 mm, SUVmax 14), multiple hypodense hepatic lesions (measuring up to 22 mm, SUVmax 5.3), and an hypermetabolic lesion on the small curvature and gastric antrum (measuring 53 × 48 mm, SUVmax 23). There was no lung, bone or central nervous system disease. Incisional biopsies of stomach and liver lesions showed pleomorphic cells, some of them with evident nucleoli, atypia, frequent figures of mitosis and bizarre mitosis, and some with nuclei with fine chromatin, with identified clefts, both with surface expression of S-100, langerin (partial) and CD1a, that were designated as Langerhans Cell (LC) Neoplasia, most likely being LC Histiocytosis with atypical findings. This biopsy was reviewed by the Memorial Sloan-Kettering Cancer Center (MSKCC). BRAF V600E mutation was not identified. Patient received a first cycle of Cytarabine due to limited access to Cladribine, and then 2 cycles of Cladribine. PET-CT reevaluation after 3 cycles of chemotherapy showed involution of hepatic nodules and stability in most cervical lymph nodes, but volumetric and hypermetabolic increase in gastric lesions (63 ×62 mm, SUVmax 29,7) and in most abdominal lymph nodes. Upper GI endoscopy revealed bleeding in a large antral tumor. A new gastric biopsy showed atypical histiocytoid cells, some pleomorphic, with immunohistochemistry expression of CD68 (weak), S100m, CD1a, p53 (aberrant pattern) and Ki67 (75%), favoring transformation to LCS. Additional PD-L1 immunostain showed strong membranous staining, with CPS 37,5. A molecular analysis performed with the MSK-HEME-IMPACT multigene panel showed 22 mutations, including *KRAS* c.183A>C; p.(Gln61His) missense mutation, VAF 30,7%. In the meantime, the patient developed severe abdominal pain, GI bleeding and anemia. He received gastric external beam radiation therapy for symptom control, for a total dose of 3,000 cGy over 20 fractions. Considering the high PD-L1 tumor expression and cases previously reported, anti PD-L1 therapy with Pembrolizumab was initiated. So far, the patient has received three cycles of immunotherapy, with clinical and laboratory improvement. A PET-CT after 3 months of completion of radiotherapy will be performed to evaluate radiological response. **Discussion and conclusion:** This case illustrates the aggressive course of LCS and the need for therapies beyond conventional chemotherapy, such as anti-PD1 therapy, to achieve disease control. Clinical trials for ultra-rare diseases are challenging to be conducted and discussion between centers may provide insightful treatment recommendations.

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LINFOMA NÃO-HODGKIN FOLICULAR NO CANAL MANDIBULAR DIREITO EM UM PACIENTE DE 25 ANOS, UM RELATO DE CASO

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Introdução/Objetivos: O linfoma folicular é a forma mais comum de Linfoma Não-Hodgkin (LNH) no Brasil, correspondendo a cerca de 20% dos casos de todos os linfomas. Esta neoplasia surge de células do centro germinativo e está intimamente associada a translocações envolvendo alterações em IgH e BCL2 nos cromossomos 14 e 18. O presente estudo visa divulgar o caso de um paciente em tratamento de LNH folicular grau 3A, com uma apresentação não-habitual, salientando a importância de uma equipe multidisciplinar bem instruída, com o objetivo de disseminar o conhecimento científico acerca desta doença. **Material e métodos:** Coleta e análise da história clínica do paciente, a partir de entrevista e exames prévios. Ademais, foram realizadas pesquisas nas bases de dados SciELO e MEDLINE, a fim de investigar e analisar os diversos aspectos acerca do LNH folicular e possíveis casos semelhantes ao relatado neste estudo. **Resultados:** Paciente masculino, 25 anos, sem doenças prévias e com avô materno diagnosticado com câncer de próstata. Procurou serviço de saúde devido ao aparecimento de pequenos e dolorosos nódulos axilares, imóveis e com secreção purulenta, sendo diagnosticado e tratado como infecção bacteriana. Após 5 meses, evoluiu, sem melhora, com o surgimento de um novo nódulo grande e indolor, na parte inferior interna direita da cavidade oral, dificultando a mastigação. Retornou ao serviço de saúde, sendo encaminhado ao odontologista, que indicou uma biópsia. Paciente relata redução dos nódulos axilares e excisão do bucal. O resultado da imunohistoquímica foi positivo para os anticorpos CD20, CD10 e BCL6 nas células neoplásicas; CD3 em linfócitos T reativos e índice proliferativo (KI67) de 80% nos folículos. Diagnosticou-se LNH folicular grau 3A, prosseguindo para o serviço médico especializado. A partir disso, foi preconizado o tratamento quimioterápico, o qual o paciente ainda realiza. **Discussão:** O linfoma folicular apresenta-se de forma característica como linfadenopatia indolor, generalizada, em adultos de meia-idade, sendo raros em pacientes mais jovens. No entanto, por mais que haja uma apresentação clássica dos linfomas, esta pode variar, com achados não-característicos, fazendo assim, necessária uma equipe multidisciplinar capacitada e engajada. No caso descrito, a alteração nodular foi observada inicialmente pelo odontologista, que encaminhou para a biópsia, permitindo um diagnóstico preciso e direcionando o plano terapêutico do paciente. **Conclusão:** A partir desta apresentação e das variáveis analisadas neste trabalho, é fato que os linfomas podem se manifestar diversamente. Logo, faz-se evidente a necessidade de um grande engajamento entre os diversos profissionais da equipe médica e também do setor odontológico, concretizando uma equipe multidisciplinar sólida e efetiva, para que diagnósticos mais precisos