

EARLY-STAGE MANTLE CELL LYMPHOMA INVOLVING THE STOMACH: SUSTAINED REMISSION AFTER ISOLATED INVOLVED-FIELD RADIOTHERAPY

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Objective: To report a case of an extremely rare form of mantle-cell lymphoma (MCL) achieving long-term remission after IF-RT. **Materials and methods:** Data were obtained through the review of clinical records. **Results:** A 57-year-old man, with history of hypertension, presented to the Emergency Room with hematemesis and abdominal pain in December 2021. The patient denied fever, adenomegaly, sweating or changes in gastrointestinal habits, and reported intentional weight loss of 10 kg in 4 months related to dietary changes. He underwent an Upper Digestive Endoscopy (UDE) upon admission, with the presence of Mallory-Weiss tear in the distal esophagus and Forrest III duodenal ulcer. Another endoscopy was performed in March 2022 and showed elevated lesions in the fundus and gastric body. Also, endoscopic ultrasonography evidenced the presence of multiple sparse elevated lesions in the gastric body, with hypoechoic echotexture, homogeneous, with precise limits, inserted in the deep mucosa and submucosa, measuring up to 35 mm x 17 mm. A biopsy of gastric lesions was done in April 2022, with positive immunohistochemistry for CD5, CD20, Cyclin D1, BCL-2 and Ki-67 (20-30%), and negative for BCL-6, CD3, MUM-1, CD10 and CD23, compatible with classical MCL. Bone marrow biopsy, aspirate and immunophenotyping from May 2022 were negative for MCL infiltration. The PET-CT from June 2022 showed increased glucose uptake in the gastric mucosa, with a diffuse and heterogeneous pattern (SUV 6.2), associated with thickening of the stomach walls, extending to the pylorus (SUV 4.6), with no evidence of other high-uptake areas. The patient was diagnosed with primary gastric MCL, stage IEA and high-risk MIPI score. Treatment with isolated IF-RT, in 24 fractions of 1.5Gy with a total dose of 36Gy, between July and August 2022, was chosen. During follow-up, the patient showed a complete response on the PET-CT from October 2022, as well as normal gastric biopsy results from September 2022 and June 2023. As of July 2023, the patient had no reported complaints and presented with normal blood count and LDH, and he was scheduled a follow-up every 3 months with periodic reassessments with UDE. **Discussion:** Primary gastric MCL is an extremely rare form of gastric tumor that accounts for less than 3% of MCLs, with no established optimal management and very poor response rates. Proposed therapeutic strategies involve observation; localized surgery or radiation therapy; less aggressive therapies with or without autologous stem-cell transplant (ASCT); and aggressive therapies with or without ASCT may be used, but the impact of enhanced systemic treatment approaches on improving outcomes remains uncertain. **Conclusion:** We reported a case of primary gastric MCL treated with exclusive IF-RT to the stomach, with a sustained complete response during the 12-month follow-up. This, together with previous

reports, implies that less aggressive frontline treatment might be justifiable for early-stage MCL involving the upper gastrointestinal tract, considering the satisfactory outcome achieved.

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HHV-8-ASSOCIATED MULTICENTRIC CASTLEMAN DISEASE IN AN HIV CARRIER COMPLICATED BY HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS AND TUMOR LYSIS SYNDROME: ATYPICAL PRESENTATIONS IN A RARE LYMPHOPROLIFERATIVE DISORDER

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Introduction/Objective: Human herpesvirus-8 (HHV-8)-associated multicentric Castleman disease (MCD) is a rare lymphoproliferative disorder sometimes associated with acquired human immunodeficiency syndrome. Hemophagocytic lymphohistiocytosis (HLH) and tumor lysis syndrome (TLS) are unusual complications of MCD. Here, we presented a case report of an HIV patient with MCD who evolved for HLH and TLS, atypical presentations in this clinical setting. **Methods:** Data were obtained reviewing the medical record of a patient diagnosed and treated at the University of São Paulo (USP) in 2023. **Case report:** A 31 year-old man was admitted at the emergency department of HC-USP in June 2023 presenting multiple adenomegalies, recurrent fever and weight loss during the past 4 months. He had a previous diagnosis of HIV infection in regular treatment, with a T-CD4 lymphocyte count = 514 copies/mL. The patient was previously admitted in another service due to infectious complications, where he underwent an axillary lymph node biopsy, which showed lymphoid hyperplasia rich in interfollicular plasmacytes, compatible with a HHV-8-associated MCD, in addition to a spindle-cell neoplastic proliferation, supporting the concomitant diagnosis of Kaposi's sarcoma (KS). He was submitted to up-front treatment based on prednisone 1 mg/kg/day. Upon admission at the HC-USP, he presented hepatosplenomegaly, ascites, and multiple lymph node enlargement. Laboratory evaluation revealed anemia, thrombocytopenia, hypoalbuminemia and marked increase in inflammatory tests, fulfilling criteria for MCD activity. After 10 days, he evolved with worsening of hepatosplenomegaly, progressive increase of direct bilirubin, pancytopenia, hyperferritinemia and hypertriglyceridemia. He also had TLS requiring dialysis. Due to this presentation, a bone marrow evaluation was performed, which showed many figures of hemophagocytosis and high EBV viral load measured by qRT-PCR. The H-Score was applied to this case, with a result of 228 points, inferring 98-99% probability of HLH diagnosis, compatible with the previously formulated hypothesis. Promptly, methylprednisolone pulsotherapy was started at a dose of 1 g/day I.V. and etoposide 50 mg/m²/day I.V. for 3 days. Despite this approach, the patient presented an unfavorable evolution, evolving to death

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, particularly 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/mm³, atypical lymphocytes and platelets 15000/mm³; 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 microvacuolization, 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/mm³ with 45% blasts and platelets 25000/mm³. 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,