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 plasmocytes, compatible with a HHV-8-associated MCD, in addition to a spindle-cell neoplastic proliferation, supporting theconcomitant 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 dyalisis. 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/m^2/day I.V. for 3 days. Despite this approach, the patient presented an unfavorable evolution, evolving to death