Primary colonic DLBCL is extraordinarily rare, often mimicking adenocarcinoma both clinically and endoscopically, potentially leading to diagnostic delays or mismanagement. The double-expressor phenotype, present in this case, represents an aggressive biological subset associated with poor prognosis and potential resistance to standard R-CHOP therapy. The absence of genetic translocations distinguished this case from double-hit lymphoma, which would have warranted even more intensive treatment approaches. However, the double-expressor status combined with extremely high Ki-67 suggests consideration of dose-adjusted EPOCH-R or other intensified regimens over standard R-CHOP. The isolated colonic presentation without nodal or bone marrow involvement represents stage I disease, potentially offering better outcomes despite the adverse biological features. **Conclusion:** Primary colonic DLBCL with double-expressor phenotype represents a rare but aggressive entity requiring prompt recognition and specialized treatment. Comprehensive immunohistochemical and molecular evaluation is essential for accurate classification and optimal therapeutic decision-making in this challenging clinical scenario.

https://doi.org/10.1016/j.htct.2025.106122

OP 13

CD20 Antigen Loss in T-cell/Histiocyte-Rich Diffuse Large B-cell Lymphoma Following R-CHOP Therapy: A Case of Immune Escape

Ayşegül Ezgi Çetin *, Ali Turunç, Berrak Çağla Şenol, Birol Güvenç

Çukurova University, Dept.of Hematology, Balcali_Adana, Turkiye

Introduction: T-cell/histiocyte-rich diffuse large B-cell lymphoma constitutes approximately 1-3% of all DLBCL cases, characterized by scattered large B-cells within an extensive reactive T-cell infiltrate. This rare variant demonstrates unique biological features including frequent immune evasion mechanisms and resistance to standard immunochemotherapy. CD20 antigen loss following rituximab-containing regimens represents a well-recognized but uncommon immune escape phenomenon, occurring in approximately 10-20% of relapsed/refractory DLBCL cases. **Case Report:** A 64-year-old female presented in late 2024 with abdominal pain, weight loss, and constitutional symptoms. Imaging studies revealed para-aortic lymphadenopathy, and trucut biopsy demonstrated T-cell/histiocyte-rich DLBCL with immunohistochemical profile showing CD20(+), PAX5(+), MUM1(+), CD3 (+) reactive T-cells, and negative CD30. The diagnosis was confirmed by the presence of scattered large B-cells within an extensive CD3+ T-cell background. The patient received in outer clinic standard R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisolone) for 6 cycles at an external center. Post-treatment PET-CT in March 2025 demonstrated complete metabolic response with no residual FDG uptake in previously involved lymph nodes (Deauville score 1-2). Unfortunately, the patient developed early relapse within 3 months, presenting in June 2025 with left cervical lymphadenopathy, constitutional symptoms, and weight loss. Restaging PET-CT revealed extensive disease with left cervical lymph nodes (22×15 mm, SUVmax: 7.96), massive para-aortic/iliac mass ($87 \times 42 \times 212$ mm, SUVmax: 7.96), left lung parenchymal involvement, and diffuse bone marrow activity. Bone marrow biopsy performed in July 2025 confirmed lymphomatous infiltration with a striking finding: complete loss of CD20 expression while maintaining PAX5(+) and MUM1(+) positivity, with persistent extensive reactive CD3+ T-cell infiltrate. This represented clear evidence of CD20 antigen loss as an immune escape mechanism following rituximab exposure. Given the patient's cardiac dysfunction precluding anthracycline-containing regimens, early relapse with CD20 negativity, and extensive disease burden and according to Turkish insurance systems low dose pralatrexate planned to targeting the T-cell-rich microenvironment in refractory settings. **Discussion:** This case illustrates several critical aspects of TCRLBCL management. The rapid relapse despite initial complete response highlights the aggressive nature of this DLBCL variant and its propensity for immune escape. The complete loss of CD20 antigen represents a well-documented resistance mechanism whereby malignant B-cells evade rituximab-mediated cytotoxicity through antigen downregulation or loss. The extensive reactive T-cell infiltrate characteristic of TCRLBCL may contribute to both immune surveillance and paradoxically provide a protective microenvironment for malignant cells. This unique tumor microenvironment necessitates novel therapeutic approaches targeting both malignant B-cells and the surrounding immune milieu. The importance of re-biopsy at relapse cannot be overstated, as demonstrated by the critical finding of CD20 loss that fundamentally altered treatment planning from CD20-targeting to alternative approaches.

https://doi.org/10.1016/j.htct.2025.106123

OP 14

Asymptomatic Waldenström Macroglobulinemia: A Case of Incidental Monoclonal Gammapathy Discovery

Naciye Nur Tozluklu, Birol Güvenç

Çukurova University, Dept.of Hematology, Balcali_Adana, Turkiye

Introduction: Waldenström macroglobulinemia is a rare B-cell malignancy representing less than 2% of all hematologic malignancies, with an annual incidence of approximately 3-5 cases per million. The disease is characterized by lymphoplasmacytic lymphoma infiltrating bone marrow and lymphoid organs with concurrent IgM monoclonal protein secretion. Many patients are diagnosed asymptomatically through incidental laboratory findings, requiring careful evaluation to distinguish from other B-cell disorders and determine appropriate management strategies. **Case Report:** A 54-year-old female was referred to hematology following discovery of a monoclonal spike on routine serum protein electrophoresis during routine health screening. The patient denied symptoms suggestive of hyperviscosity syndrome including headache, visual disturbances, epistaxis, or neurological

complaints. She reported no B-symptoms (fever, night sweats, weight loss) and had no history of recurrent infections or bleeding tendencies. Physical examination was unremarkable without palpable lymphadenopathy, hepatomegaly, or splenomegaly. The patient appeared well with stable vital signs and no evidence of hyperviscosity syndrome. Laboratory evaluation revealed significant findings on protein studies. Serum protein electrophoresis showed increased gamma fraction (26.3%; normal: 10.7-20.3%) with relatively decreased albumin (47.9%; normal: 52-65%) and albumin/globulin ratio of 0.92. A sharp M-spike was evident in the gamma region. Immunofixation electrophoresis confirmed IgM-kappa monoclonal protein. Quantitative immunoglobulins demonstrated markedly elevated IgM at 27.98 g/L with normal IgG (7.6 g/L) and IgA (2.4 g/L). Beta-2 microglobulin was normal (1.91 mg/L), indicating low tumor burden. Urine free light chain analysis showed normal kappa (3.78 mg/L) and lambda (0.73 mg/L) levels with elevated kappa/lambda ratio (5.18), consistent with kappa-predominant monoclonality. Bone marrow examination revealed 40% cellularity with approximately 25% infiltration by small B-lymphocytes with plasmacytic differentiation organized in 4-5 intertrabecular lymphoid aggregates. Reticulin fibrosis was absent (grade 0/4), and amyloid staining was negative. Immunohistochemistry demonstrated CD20+, CD38+, CD138+ cells with negative CD5, CD23, cyclin D1, LEF-1, and CD56, excluding chronic lymphocytic leukemia/small lymphocytic lymphoma and mantle cell lymphoma. Flow cytometry confirmed CD19+/CD20+/CD45+ clonal B-cell population with CD138+ plasmacytic subset showing intracytoplasmic kappa restriction and negative CD56, consistent with lymphoplasmacytic lymphoma rather than multiple myeloma. Based on the constellation of findings including IgM-kappa monoclonal protein, characteristic bone marrow morphology and immunophenotype, the diagnosis of lymphoplasmacytic lymphoma/Waldenström macroglobulinemia was established. **Discussion:** This case illustrates typical presentation of asymptomatic WM discovered through routine screening. The markedly elevated IgM level (27.98 g/L) without hyperviscosity symptoms demonstrates the variable clinical presentation of WM patients. The characteristic immunophenotype (CD20+/CD38+/CD138+/CD5-/CD23-/CD56-) with intracytoplasmic kappa restriction distinguishes WM from other B-cell disorders. Current management guidelines recommend "watch and wait" approach for asymptomatic WM patients without end-organ damage or symptomatic disease. However, given the markedly elevated IgM level, careful monitoring for hyperviscosity syndrome development is essential. Molecular testing for MYD88 L265P mutation (present in >90% of WM cases) would provide diagnostic confirmation and prognostic information regarding treatment response, particularly to BTK inhibitors. **Conclusion:** Asymptomatic Waldenström macroglobulinemia requires comprehensive diagnostic evaluation to confirm diagnosis and assess disease burden. Despite markedly elevated IgM levels, many patients can be safely observed with regular monitoring, emphasizing the importance of individualized management approaches in this rare but well-characterized lymphoproliferative disorder.

https://doi.org/10.1016/j.htct.2025.106124

OP 15

Primary Extranodal Marginal Zone Lymphoma of the Maxilla with Sphenoid Bone Invasion: Excellent Response to R-CHOP Therapy

Berrak Çağla Şenol*, Ayşegül Ezgi Çetin, Ali Turunç, Birol Güvenç

Çukurova University, Dept.of Hematology, Balcali_Adana, Turkiye

Case Report: A 59-year-old male (weight: 65 kg, height: 168 cm) presented in September 2024 with a progressively enlarging mass in the right maxillary region extending toward the temporal area with sphenoid bone proximity. The patient complained of maxillary distortion and pain but denied B symptoms including fever, night sweats, or weight loss. Physical examination revealed facial asymmetry with palpable right maxillary swelling. Initial biopsy of the maxillary mass demonstrated CD20-positive extranodal marginal zone lymphoma consistent with MALT lymphoma histology. Staging F-18 FDG PET/CT performed on October 4, 2024, revealed a hyperintense soft tissue mass in the right maxillary region with sphenoid bone invasion showing SUVmax 6.81. Additionally, a 16 × 10 mm lymph node in the right level 2 cervical chain demonstrated SUVmax 4.22. No pathological FDG uptake was detected in the thorax, abdomen, or skeletal system, confirming localized disease. Based on the diagnosis of localized EMZL with bone invasion and cervical lymph node involvement, standard R-CHOP chemotherapy was initiated on September 24, 2024. The regimen consisted of rituximab 375 mg/m^2 (day 1), cyclophosphamide 750 mg/m^2 (day 1), doxorubicin 50 mg/m^2 (day 1), vincristine 1.4 mg/m^2 (day 1), and prednisolone 100 mg daily for 5 days. Supportive care included G-CSF (filgrastim) for neutropenia prophylaxis and antiemetics (ondansetron, granisetron). During treatment, the patient developed E. coli pneumonia, which resolved with appropriate antibiotic therapy and supportive care. Despite this complication, the treatment protocol was successfully completed. Interim PET-CT evaluation on December 25, 2024, demonstrated significant metabolic response with maxillary lesion SUVmax decreasing from 6.81 to 2.92, accompanied by dimensional reduction. Cervical lymph node involvement was no longer detectable, yielding a Deauville score of 2, consistent with partial metabolic remission. Follow-up PET-CT after completion of 4 cycles in March 2025 revealed complete disappearance of pathological FDG uptake throughout the body. The maxillary region showed no residual mass formation, maintaining Deauville score 2, confirming complete metabolic remission. The patient tolerated treatment well overall and entered surveillance follow-up without evidence of systemic dissemination or bone marrow involvement. **Discussion:** This case represents a rare presentation of EMZL involving the maxillofacial region with sphenoid bone invasion. The excellent response to standard R-CHOP therapy challenges the traditional approach of radiotherapy alone for localized EMZL, particularly in cases with bone involvement where complete surgical resection may not be feasible. The use of PET-CT for treatment response assessment proved invaluable, providing objective metabolic parameters through