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.

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OP 15

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

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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

Deauville scoring system. The dramatic reduction in SUVmax values (from 6.81 to undetectable levels) correlated with excellent clinical response. EMZL typically follows an indolent course with favorable prognosis. However, bone involvement may indicate more aggressive behavior, potentially justifying systemic chemotherapy over local treatments. The complete metabolic remission achieved in this case supports the efficacy of R-CHOP in this clinical scenario. **Conclusion:** Primary EMZL of the maxilla with sphenoid bone invasion represents a rare clinical entity that can be successfully treated with standard R-CHOP chemotherapy. PET-CT monitoring using Deauville scoring provides valuable objective assessment of treatment response. This case contributes to the limited literature on optimal management of localized EMZL with bone involvement.

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OP 16

Double-Expressor Diffuse Large B-Cell Lymphoma of Bone and Soft Tissue in a 29-Year-Old Patient: A Rare Case Report

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Case report: Primary bone lymphoma represents less than 1% of all malignant bone tumors and approximately 3% of extranodal lymphomas. Diffuse large B-cell lymphoma constitutes the most common histological subtype of primary bone lymphoma, typically affecting adults with a slight male predominance. The clinical presentation often mimics primary bone sarcomas, potentially leading to diagnostic delays. Double-expressor lymphomas, characterized by MYC and BCL2 protein co-expression, constitute 20-30% of DLBCL cases and are associated with inferior outcomes compared to standard DLBCL, requiring consideration of intensified treatment regimens. A 29-year-old male presented with several months of progressive upper extremity pain, swelling, and limited range of motion involving the long bones and scapula. The clinical presentation initially raised suspicion for osteosarcoma or soft tissue sarcoma, prompting orthopedic evaluation and excisional biopsy. Macroscopic examination revealed approximately 4 cm of grayish-white to brown tissue fragments submitted for histopathological analysis. Microscopic evaluation demonstrated cellular morphology consistent with lymphoproliferative disease rather than sarcomatous features, prompting comprehensive immunohistochemical evaluation. Immunohistochemical analysis confirmed lymphoid origin with positive LCA (leukocyte common antigen) staining. B-cell lineage was established by strong, diffuse CD20 positivity (80-85% of cells). The tumor demonstrated germinal center B-cell phenotype with BCL6 expression in 80-85% of cells. Critically, MYC expression was present in 40-45% of tumor cells, suggesting double-expressor status pending BCL2 confirmation. The proliferation index was extremely high with Ki-67 staining positive in 80-85% of cells, indicating highly aggressive biology. Negative staining for MyoD1, CD34, S100, and CD3 excluded sarcomatous differentiation and T-cell lymphoma. Based on the constellation of findings, the diagnosis of high-grade diffuse large B-cell lymphoma with germinal center phenotype and suspected double-expressor features was established. The anatomical location involving upper extremity long bones and scapula confirmed primary bone lymphoma classification. Additional molecular studies were recommended including FISH analysis for MYC, BCL2, and BCL6 rearrangements to distinguish between double-expressor and double-hit lymphoma. Comprehensive next-generation sequencing panel evaluation was suggested focusing on prognostically relevant genes including TP53, CDKN2A/B, NOTCH1/2, EZH2, and other lymphoma-associated mutations. **Discussion:** This case illustrates several important clinical and pathological considerations. Primary bone DLBCL in young adults is uncommon and may present diagnostic challenges due to clinical similarity to primary bone sarcomas. The initial clinical suspicion of sarcoma necessitated careful immunohistochemical evaluation to establish correct diagnosis. The double-expressor phenotype with MYC and suspected BCL2 co-expression, combined with extremely high Ki-67 proliferation index (80-85%), indicates aggressive biology requiring intensive treatment approaches. While confirmation of BCL2 expression and FISH analysis for genetic rearrangements remain pending, the current findings suggest consideration of dose-adjusted EPOCH-R or similar intensified regimens rather than standard R-CHOP therapy. The young age of the patient and localized bone involvement may offer favorable prognostic factors despite the aggressive biological features. However, the high proliferation index and suspected double-expressor status necessitate careful treatment planning with multidisciplinary input. **Conclusion:** Primary bone DLBCL with double-expressor features in young adults represents a rare but aggressive entity requiring prompt recognition and intensive treatment. This case emphasizes the importance of comprehensive immunohistochemical evaluation in suspected bone malignancies and highlights the need for molecular characterization to guide optimal therapeutic approaches in high-grade B-cell lymphomas.

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Myeloma

OP 17

Familial Multiple Myeloma in a Post-Renal Transplant Patient: A Case of Smoldering Multiple Myeloma with Strong Family History

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Introduction: Familial multiple myeloma represents approximately 1-2% of all MM cases, characterized by the occurrence of MM in two or more first-degree relatives. While the exact genetic mechanisms remain unclear, several familial