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

clustering studies suggest inherited susceptibility genes and shared environmental factors. Immunosuppression following solid organ transplantation may accelerate malignant transformation in genetically predisposed individuals, creating a unique clinical scenario requiring specialized monitoring and management approaches. **Case Report:** A 50-year-old female with a complex medical history presented with fatigue, weakness, and anemia. Her medical background included type 1 diabetes mellitus diagnosed in 1982 at age 8, progression to end-stage renal disease secondary to diabetic nephropathy in 2001, and successful deceased donor kidney transplantation in 2007. She remained on chronic immunosuppressive therapy with mycophenolic acid (Myfortic®) and cyclosporine (Sandimmun®) with stable graft function. The patient's family history was remarkable for multiple myeloma: her mother was alive with confirmed MM diagnosis, and her brother had previously died from MM after receiving treatment. This strong familial clustering placed her in the high-risk category for hereditary MM predisposition. Physical examination revealed pallor consistent with anemia, but no lymphadenopathy, bone tenderness, or other significant findings. Laboratory evaluation demonstrated significant anemia (hemoglobin 7.8 g/dL, hematocrit 26.2%) with normocytic indices (MCV 87 fL). Renal function remained stable post-transplant, and serum calcium was within normal limits. Protein studies revealed elevated beta-2 fraction on serum protein electrophoresis with positive IgG-kappa monoclonal band on immunofixation electrophoresis. Free light chain analysis showed elevated kappa (40.7 mg/L) with kappa/lambda ratio of 1.86. Bone marrow examination demonstrated 3-4% plasma cells with flow cytometry confirming CD138+/CD38+ phenotype and kappa light chain restriction (80% kappa, 20% lambda), establishing clonality. Comprehensive FISH analysis was negative for high-risk cytogenetic abnormalities including p53 deletion, del(13q), t(11;14), and t(4;14). Lumbar MRI revealed disc protrusions without lytic bone lesions. Genetic analysis for FMF mutations was performed given potential inflammatory contributions, showing R202Q heterozygosity and other polymorphisms without pathogenic significance. Based on the presence of IgG-kappa monoclonal protein, 3-4% clonal bone marrow plasma cells, anemia, and absence of hypercalcemia or lytic lesions, the patient was diagnosed with smoldering multiple myeloma. **Discussion:** This case illustrates several important aspects of familial MM. The strong family history with both maternal and sibling involvement suggests significant genetic predisposition, warranting enhanced surveillance protocols. The co-existence of chronic immunosuppression following renal transplantation creates additional complexity, as immunosuppressive agents may accelerate progression from precursor states to overt malignancy.

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OP 18

CD56-Negative IgA-Lambda Multiple Myeloma with Bortezomib-Induced Severe Cutaneous Reaction: A Case Report

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We report a 51-year-old male with IgA-lambda multiple myeloma who developed severe cutaneous drug eruption following bortezomib treatment. Despite treatment modification to daratumumab-based regimen, the patient achieved complete remission, demonstrating successful management of therapy-related adverse events in CD56-negative myeloma phenotype. Introduction: Multiple myeloma represents approximately 10% of hematologic malignancies, with CD56-negative variants comprising a rare subset associated with distinct clinical characteristics. Bortezomib-containing regimens remain first-line therapy; however, cutaneous adverse reactions can necessitate treatment modifications. We present a case of successful alternative therapy following severe bortezomib-induced skin toxicity. Methods/Case Presentation: A 51-year-old male presented with fatigue and back pain. Laboratory investigations revealed IgA elevation (6.8 g/L) with lambda light chain restriction. Serum protein electrophoresis showed decreased albumin (51.6%) and elevated beta fractions. Bone marrow flow cytometry demonstrated plasma cell population: CD38/CD138 100%, CD45 100%, CD117 79.8%, CD56 7.5% (negative), with 96.7% lambda clonality, confirming IgA-lambda multiple myeloma with CD56-negative phenotype. Staging revealed elevated β2-microglobulin (2.75 mg/L). PET/CT identified metabolically active lytic lesions in T3 vertebra (SUVmax 6.35) and right lumbosacral region (SUVmax 13.41), indicating metabolic progression without hepatosplenomegaly. Initial treatment commenced with VRD (bortezomib, lenalidomide, dexamethasone). After cycle 1, mild erythematous pruritic rash appeared. Following cycle 2, extensive cutaneous eruptions developed. Skin biopsy revealed upper dermal eosinophil-associated perivascular infiltration with erythrocyte extravasation; direct immunofluorescence was negative, consistent with drug-induced eruption. Bortezomib was discontinued, and treatment switched to DRd (daratumumab, lenalidomide, dexamethasone). After 2 DRd cycles, M-protein disappeared, serum and urine immunofixation became negative, and hematologic parameters normalized. Follow-up PET/CT showed no active myeloma lesions, confirming complete remission. Results: The patient achieved biochemical and radiological complete remission within 2 cycles of daratumumab-based therapy following bortezomib-induced severe cutaneous reaction. No significant toxicities were observed with the modified regimen. Discussion: CD56-negative multiple myeloma represents a rare