Lymphoma

OP 8

VITREORETINAL INVOLVEMENT IN NASAL CAVITY B-CELL LYMPHOMA: A RARE FORM OF RELAPSE

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INTRODUCTION: Non-Hodgkin lymphomas are malignant neoplasms of lymphoid tissue, and a subset present with extranodal involvement. The head and neck region represents one of the clinically relevant localizations. Sinonasal B-cell lymphomas are a rare subtype, most often manifesting as diffuse large B-cell lymphoma (DLBCL), and typically show aggressive clinical behavior. Relapses most frequently involve cervical lymph nodes, the orbit, and the central nervous system. Ocular involvement is rare, usually presenting as orbital masses or ocular adnexal lymphoma. Vitreoretinal infiltration is even more unusual and has been described only infrequently. In this case report, we present an elderly male patient with nasal cavity B-cell lymphoma who developed relapse with vitreoretinal involvement, aiming to emphasize the diagnostic and therapeutic aspects of this rare condition. **CASE PRESENTATION:** A 71-year-old male was diagnosed three years earlier with nasal cavity B-cell lymphoma. Bone marrow biopsy at diagnosis showed no systemic involvement. He received four cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) and achieved complete remission. Three years later, he presented with decreased vision in the left eye. Orbital MRI showed tortuosity of the optic nerve and slight widening of the perioptic space (Figure 1). Cranial MRI revealed only age-related changes. Cytology and flow cytometry of vitreous fluid demonstrated CD20 and CD79a positivity with high proliferative activity, consistent with B-cell neoplasia. PET-CT revealed limited FDG uptake (SUVmax 5.02) in the anterior aspect of the left orbit (Figure 2), with no additional systemic involvement. Based on his disease history, systemic high-dose methotrexate combined with cytarabine and intrathecal therapy was initiated. Radiotherapy was also considered. He was referred to another specialized center for possible intravitreal chemotherapy. Despite systemic treatment, follow-up revealed that the patient had died. **DISCUSSION AND CONCLUSION:** Sinonasal B-cell lymphomas are uncommon, most often exhibiting DLBCL histology with aggressive clinical features. Relapses most frequently involve cervical nodes, orbital structures, or the central nervous system. Although orbital disease is recognized, vitreoretinal infiltration is exceedingly rare and has been reported in less than 5% of cases in large series. Diagnosis is challenging, as ocular involvement may present with non-specific symptoms such as visual impairment or vitreous opacities, requiring cytology, immunophenotyping, and immunohistochemistry of vitreous samples for confirmation. Therapeutic options include systemic high-dose methotrexate and cytarabine, with intrathecal therapy commonly added for central nervous system prophylaxis. Radiotherapy may contribute to local control in orbital disease. Intravitreal chemotherapy has also been described, most often with methotrexate, and rituximab has been used in selected cases. The prognosis of ocular involvement is poor, with median survival reported between 12 and 36 months and a high risk of central nervous system relapse. This case illustrates that vitreous infiltration may represent a relapse manifestation of sinonasal B-cell lymphoma and highlights the importance of careful evaluation of ocular symptoms in such patients.

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OP 9

THERAPEUTIC CHALLENGE IN HISTIOCYTIC SARCOMA: A CASE REPORT OF NIVOLUMAB ADDITION TO THE ICE PROTOCOL

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Introduction: Histiocytic sarcoma (HS) is an exceptionally rare and aggressive hematopoietic malignancy, representing less than 1% of hematologic neoplasms [1]. No standardized therapeutic regimen exists; patients are often treated with lymphoma-like regimens such as CHOP or ICE, with limited efficacy and median survival of approximately six months [2,1]. Recent advances in molecular pathology have revealed recurrent BRAF^V600E mutations, ALK rearrangements, and PD-L1 expression, providing new diagnostic and therapeutic implications [3]. Case-based evidence suggests that PD-1 inhibitors may induce durable responses in select patients with PD-L1–positive HS [4,5]. **Case Presentation:** A 28-year-old male presented with abdominal pain and swelling. Imaging demonstrated a large intra-abdominal mass with peritoneal implants. Histopathology confirmed HS, positive for CD45, CD163, and CD14, with a Ki-67 index of 80%. Bone marrow biopsy was normocellular. Molecular analysis excluded BRAF and ALK alterations but demonstrated PD-L1 expression with a tumor proportion score (TPS) of 1–49% and a combined positive score (CPS) of 35%. The patient was started on ICE chemotherapy (ifosfamide, carboplatin, etoposide). Following biomarker analysis, nivolumab was introduced beginning with the second cycle. The treatment was well tolerated, and subsequent PET-CT demonstrated marked metabolic regression with clinical improvement. Follow-up abdominal imaging confirmed complete radiological response, with disappearance of the initially described mesenteric mass. **Conclusion:** Discussion HS poses a therapeutic challenge because of its aggressive course and lack of standardized therapy [2,1]. Conventional chemotherapy regimens have limited durability, and reported responses are often transient. The presence of PD-L1 expression provided a rationale for incorporating a PD-1 inhibitor, even at moderate expression levels, consistent with emerging literature [4]. Previous case reports have demonstrated clinical benefit from

pembrolizumab and nivolumab in PD-L1–positive HS, including durable complete responses [5]. In this patient, radiological assessment corroborated complete remission after combined ICE and nivolumab, supporting the potential role of checkpoint inhibition in improving depth of response. This case represents one of the few documented examples of combining intensive chemotherapy with checkpoint blockade in HS, highlighting the potential synergistic role of immunotherapy.Conclusion This case illustrates the rarity and therapeutic complexity of HS. The addition of nivolumab to ICE chemotherapy, guided by PD-L1 expression, resulted in meaningful clinical response in a young patient with advanced disease. These findings underscore the importance of integrated histopathological and molecular assessment in guiding personalized management for HS.Keywords: Histiocytic sarcoma; Nivolumab; ICE protocol; PD-L1; ImmunotherapyReferences1.Takimoto, T., et al. (2023). Histiocytic sarcoma: A clinicopathologic analysis of 50 cases. American Journal of Surgical Pathology, 47(1), 1–12. 2. Emile, J. F., et al. (2022). Histiocytic and dendritic cell neoplasms: Update of the 2022 WHO classification. Blood, 140(11), 1200–1218. 3. Go, H., et al. (2019). Frequent detection of BRAF V600E mutations in histiocytic and dendritic cell neoplasms. Histopathology, 74(3), 389–400. 4. Bossard, C., et al. (2021). PD-1/PD-L1 blockade in rare hematologic malignancies: Case reports and literature review. Hematological Oncology, 39(3), 327–334. 5.Yoon, D. H., et al. (2022). Efficacy of pembrolizumab in histiocytic sarcoma with high PD-L1 expression: Case report and review. Annals of Hematology, 101(7), 1525–1530.

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OP 10

Plasma-Cell–Predominant Idiopathic Multicentric Castleman Disease: A Rare Diagnostic and Therapeutic Challenge

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Introduction: Castleman disease represents a rare, heterogeneous group of lymphoproliferative disorders, often categorized as unicentric or multicentric, with plasma-cell (PC), hyaline-vascular, or mixed histology. Idiopathic multicentric Castleman disease (iMCD) remains a diagnostic and therapeutic challenge, particularly in patients presenting with systemic inflammation and polyclonal plasmacytosis without overt clonal plasma cell disorder. We present the case of a patient with plasma-cell–predominant iMCD, successfully treated with IL-6 blockade, emphasizing the diagnostic pitfalls and the importance of early therapeutic intervention. **Methods:** A male patient was admitted to the Department of Hematology, Çukurova University, with a 1-year history of progressive fatigue, weight loss, abdominal fullness, and generalized lymphadenopathy. Physical examination revealed widespread lymphadenopathy and splenomegaly. Laboratory tests demonstrated normocytic anemia, elevated CRP and ferritin, mildly increased IgG, and elevated β2-microglobulin. Excisional lymph node biopsy and splenectomy specimens were evaluated by histopathology and immunohistochemistry. Imaging studies included CT and PET-CT for staging., Türkiye **Results:** Histopathology revealed follicular hyperplasia with regressed germinal centers and interfollicular plasmacytosis. Immunohistochemistry confirmed CD38+ and CD138+ plasma-cell infiltration, HHV-8 negativity, and a nonclonal kappa/lambda pattern. IgG4/IgG ratio was 22%. PET-CT demonstrated widespread FDG-avid lymphadenopathy (SUVmax 4–6) and splenomegaly, without extranodal organ involvement. Bone marrow evaluation was negative for clonal plasma cell infiltration. The case was classified as idiopathic multicentric Castleman disease, plasma-cell variant (iMCD-PC). The patient was initiated on tocilizumab (anti-IL-6R) in combination with corticosteroids. Within 6 weeks, systemic symptoms and inflammatory markers improved significantly, with partial regression of lymphadenopathy on imaging. In the event of refractoriness, lenalidomide or sirolimus were considered as second-line options. Close follow-up with PET-CT and serum paraproteins was arranged to monitor potential clonal evolution into plasma cell neoplasia. **Discussion:** This case illustrates the diagnostic complexity of iMCD-PC, which may mimic lymphoid malignancies and overlap with monoclonal gammopathies. The absence of monoclonality and CRAB criteria excluded multiple myeloma, while systemic inflammatory features and IL-6 axis dysregulation supported iMCD. Tocilizumab provided meaningful clinical and biochemical improvement. The case is valuable as an example of iMCD with strong plasmacytic component, highlighting the necessity of long-term surveillance due to the risk of clonal transformation. **Conclusion:** Plasma-cell–predominant iMCD is a rare and diagnostically challenging entity requiring integration of histopathology, immunohistochemistry, imaging, and laboratory findings. Anti–IL-6–directed therapy represents an effective treatment option, but close monitoring remains mandatory. This case underlines the importance of early recognition and targeted therapy in preventing disease-related morbidity.

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OP 11

LANGERHANS CELL HISTIOCYTOSIS: SINGLE-CENTER EXPERIENCE

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Introduction and Objective: Langerhans cell histiocytosis (LCH) is a rare clonal proliferative disease that can involve one or more organs (1). In adults, multisystem involvement is generally predominant (68.6%), whereas single-system involvement is less common (2). The clinical spectrum is broad, with bone, skin, and lungs being the most frequently affected organs. The treatment approach varies according to the extent of the disease, and the optimal treatment strategy