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

has not yet been clearly defined(3-6). This study aimed to evaluate the demographic characteristics, sites of involvement, treatments administered, and treatment responses of adult LCH cases diagnosed at our center. **Methods:** Medical records of adult patients diagnosed with LCH at our center between 2002 and 2024 were retrospectively reviewed. Patient age, sex, sites of involvement, treatment regimens, treatment responses, and follow-up durations were recorded. **Results:** A total of 10 patients (9 male, 1 female) were analyzed. The median age was 31.5 years (range: 20–76). The median follow-up duration was 5.8 years (approximately 69 months). Three patients (30%) had multisystem involvement, and seven patients (70%) had single-system involvement. The most common site of involvement was bone (80%), followed by skin (20%) and lymph nodes (10%). Diabetes insipidus was detected in one patient (10%). Treatment approaches were heterogeneous. Five patients received radiotherapy (RT), three patients were treated with a vinblastine and prednisolone combination, one patient with multisystem involvement received cladribine combined with RT, one patient was given prednisolone monotherapy, and one patient was followed without treatment. A response was achieved in all patients after initial treatment. Two patients (20%) experienced relapse, both in those with bone involvement only. The patient treated with cladribine remains in long-term complete remission. No mortality was observed. Feature Value Total number of patients 10 Median age (years) 31.5 (20–76) Median follow-up duration 5.8 years (approximately 69 months) Male/Female 9/1 Multisystem 3 (30%) Single-system 7 (70%) Most common involvement Bone (80%) Relapse 2 (20%) Mortality 0 **Discussion:** In adult Langerhans cell histiocytosis, multisystem involvement is reported as the most common form in the literature; however, in our study, single-system involvement was detected in 70% of patients. This discrepancy may be explained by differences in patient referral patterns to our center, follow-up of pulmonary LCH cases in chest disease clinics, variations in staging due to the retrospective design, and demographic factors.The complete remission rate with vinblastine and prednisolone combination therapy is reported to be approximately 70% in the literature (6). In our series, all three patients treated with this regimen achieved complete remission. Cladribine, a purine analog, is an effective option in refractory or relapsed cases; in the literature, monotherapy with cladribine has been reported to achieve a complete remission rate of approximately 50% and an overall response rate of approximately 90% (5). In our series, the patient treated with cladribine achieved long-term complete remission. The relapse rate in our study was 20%, consistent with the 20–30% range reported by Néel et al. (5). **Conclusion:** Although Langerhans cell histiocytosis is a rare disease, long-term complete remission can be achieved with appropriate treatment. In our study, all patients achieved a response, and the relapse rate was 20%, consistent with the literature. Multisystem involvement is a risk factor for relapse. The patient treated with cladribine achieved long-term complete remission. Larger, multicenter prospective studies are needed to optimize treatment strategies in Langerhans cell histiocytosis.

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OP 12

Primary Colonic Diffuse Large B-Cell Lymphoma with Double-Expressor Phenotype: A Rare Presentation Mimicking Adenocarcinoma

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Introduction: Primary gastrointestinal lymphomas account for approximately 1-4% of all gastrointestinal malignancies, with the colon being the least commonly affected site. Diffuse large B-cell lymphoma represents the most frequent histological subtype, but primary colonic involvement remains exceptionally rare. Double-expressor lymphomas, characterized by MYC and BCL2 protein co-expression without underlying genetic translocations, constitute 20-30% of DLBCL cases and are associated with inferior outcomes compared to standard DLBCL. The rarity of primary colonic DLBCL combined with double-expressor phenotype presents unique diagnostic and therapeutic challenges. **Case Report:** A 59-year-old male with no significant medical history presented with a 2-month history of progressive right lower quadrant abdominal pain, anorexia, and 5 kg weight loss. The patient denied fever, night sweats, or B-symptoms. Physical examination revealed mild right lower quadrant tenderness without palpable lymphadenopathy, hepatosplenomegaly, or other abnormalities. Laboratory evaluation demonstrated mild normocytic anemia (hemoglobin 11.2 g/dL) with normal leukocyte and platelet counts. Biochemical studies showed elevated lactate dehydrogenase (560 U/L) with normal renal and hepatic function. Infectious disease screening including HIV, hepatitis B, and hepatitis C serologies were negative. Computed tomography of the abdomen revealed a heterogeneous 6-cm mass involving the ascending colon wall without regional lymphadenopathy or hepatosplenic involvement. Colonoscopy identified an ulcero-vegetative mass in the ascending colon causing luminal narrowing, initially suspected to represent adenocarcinoma. Histopathological examination of colonoscopic biopsies revealed diffuse proliferation of medium-to-large sized atypical lymphoid cells with prominent nuclear atypia and high mitotic activity. Comprehensive immunohistochemical analysis demonstrated strong CD20 positivity with focal CD10 expression and positive MUM1, consistent with germinal center B-cell origin. Critical findings included diffuse BCL2 positivity and MYC expression in 70% of cells, establishing double-expressor status. The proliferation index (Ki-67) was extremely high at approximately 90%. CD3 and CD5 were negative, excluding T-cell lymphoma. Fluorescence in situ hybridization (FISH) analysis for MYC, BCL2, and BCL6 gene translocations was negative, ruling out double-hit lymphoma and confirming the diagnosis as double-expressor DLBCL rather than high-grade B-cell lymphoma with MYC and BCL2 rearrangements. The final diagnosis was primary colonic diffuse large B-cell lymphoma, germinal center subtype, with double-expressor phenotype (MYC+/BCL2+) and extremely high proliferative activity. **Discussion:** This case illustrates several important clinical and pathological considerations.