

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.

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.

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.

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OP 13

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

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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 × 42 × 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.

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OP 14

Asymptomatic Waldenström Macroglobulinemia: A Case of Incidental Monoclonal Gammopathy Discovery

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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 asymptotically 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