heterogeneous clinical presentation of FV deficiency. This supports the hypothesis that FV deficiency, due to its dual roles, may predispose not only to bleeding but also to thrombosis. Although the relationship between FV deficiency and thrombosis has not been fully elucidated, some potential mechanisms have been suggested. First, reduced TFPIα levels in patients with FV deficiency may disrupt the hemostatic balance, increasing the risk of thrombosis. Additionally, APC resistance observed in these patients could explain the hypercoagulability associated with FV deficiency. **Conclusion:** This case is significant in that it highlights the diagnosis of a rare Factor V deficiency in a patient presenting with deep vein thrombosis. While FV deficiency is typically characterized by a bleeding tendency, the predominance of a thrombotic complication in this case underscores the heterogeneous clinical course of this disorder and the challenges in its management. Although the development of thrombosis in patients with FV deficiency is rare, such cases emphasize the complexity of hemostatic balance and the necessity for individualized treatment approaches. Further case reports and a better understanding of the underlying mechanisms may lead to the development of more effective strategies for managing patients with FV deficiency.

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Adult Hematology Abstract Categories

Lymphoma
OP 04

EVALUATION OF THE IMPACT OF DEMOGRAPHIC DATA AND CLINICOPATHOLOGICAL RISK FACTORS ON TREATMENT RESPONSE AND SURVIVAL IN PATIENTS WITH PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA: A SINGLE-CENTER EXPERIENCE

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Objective: Primary central nervous system lymphoma (PCNSL) is a large B-cell lymphoma originating from specific regions such as the brain, spinal cord, and leptomeninges (1). The pathophysiology of PCNSL is not fully understood, but alterations in genes involved in the binding of immunoglobulins and B-cell receptors expressed in the central nervous system (CNS) and in NF-κB signaling are thought to play a central role (2,3). Other factors, such as T cells, macrophages/microglia, endothelial cells, chemokines, and interleukins, may also have important roles. The presenting symptoms of patients vary depending on the affected regions of the CNS (4). Standard treatment includes methotrexate-based polychemotherapy followed by age-adapted cytotoxic therapies and autologous stem cell transplantation (HDC-ASCT) (5,6). For patients ineligible for or unwilling to undergo transplantation, whole-brain radiotherapy (WBRT) or single-agent therapy may be used for consolidation (7). Personalized treatments, primary radiotherapy, or supportive care are options for patients with insufficient performance for treatment (8). Despite these treatments, 15-25% of patients do not respond to chemotherapy, and 25-50% relapse after an initial response. Although recurrence rates are higher in elderly patients, the prognosis of relapsed patients is poor, regardless of age (9). There is a need for further research to identify diagnostic biomarkers, develop more effective and less neurotoxic treatments, improve drug penetration into the CNS, and explore immunotherapies and adaptive cell therapies. **Methodology:** In this study, we aimed to evaluate the treatment responses and survival of patients with primary central nervous system lymphoma treated and followed at the Çukurova University Medical Oncology Clinic, based on their demographic data and clinicopathological risk factors. We reviewed the medical records of all lymphoma patients treated at our hospital, and 26 patients with central nervous system lymphoma were included in the study. We assessed the patients' age, gender, comorbidities, diagnosis dates, tumor locations, presenting symptoms, etiopathological factors, laboratory values, including baseline B2-microglobulin, IPI, and MSKCC scores at presentation. Pathology reports were evaluated for c-myc, Bcl-2, Bcl-6, and Ki-67 immunohistochemical markers, and FISH results, if available. The primary treatment options, treatment initiation dates, number of treatment cycles, best response, and its date were recorded. We also evaluated the radiotherapies received in addition to systemic chemotherapy. Disease progression, the date of progression, second-line treatments in case of progression, responses, and any autologous transplantation or maintenance therapies were documented. Survival outcomes, including progression-free survival and overall survival, were analyzed in relation to clinicopathological risk factors. **Results:** A total of 26 patients, 13 (50%) female and 13 (50%) male, were included in the study. The mean age was 56 (min 26- max 90). The most common comorbidity was hypertension. Regarding tumor localization, 8 patients had frontal lobe involvement, the most frequent site. The most common presenting symptoms were headache and seizures. At diagnosis, all patients required steroids, and 11 patients required antiepileptic drugs, while only 2 patients did not. Histopathological verification was performed in all patients, with all cases reported as diffuse large B-cell lymphoma. Only 1 patient had a history of hepatitis C as an etiological factor. Based on MSKCC scores at diagnosis, 8 patients were high-risk (score 3), 8 were intermediate-risk (score 2), and 8 were low-risk (score 1). Immunohistochemical analysis showed that c-myc was overexpressed in 10 patients (above 40%). Bcl-2 was positive in 12 patients, and Bcl-6 was positive in 19 patients. Five patients were triple expressors, and five were double expressors. Ki-67 ranged from 25 to 100, with 21 patients showing Ki-67 levels above 70% and 3 patients with levels above 95%. Pre-treatment laboratory tests revealed normal albumin levels in only 4 patients, while the majority had hypoalbuminemia. LDH levels were above the upper limit in

16 patients, and 3 patients had levels three times the normal limit. Inflammatory markers, including CRP and ESR, were elevated in 17 patients. The most commonly used first-line treatments were high-dose methotrexate with rituximab in 6 patients, methotrexate-rituximab-temozolomide (MTR) in 4 patients, and the MATRix protocol (methotrexate, cytarabine, thiotepa) in 3 patients. Radiotherapy, including whole-brain irradiation, was administered in 8 patients during or after first-line treatment. Ki-67 reached statistical significance when evaluating overall survival and progression-free survival with clinicopathological risk factors. No statistical significance was found for other pathological and clinical laboratory values. **Conclusion:** CNS lymphoma is a rare type of cancer that develops in central nervous system (CNS). PCNSL comprises ~4% of all primary CNS tumors. There are only about 1,500 new cases of CNS lymphoma diagnosed in each year. It occurs most often in people over 50, with the average age of diagnosis being 65. In our study, the average age of our patients was younger than the literature, and our average age was 56. It's slightly more common in men. People with HIV or AIDS and other immunodeficiencies are more likely to receive a CNS lymphoma diagnosis. Symptoms depend on where your tumor is located. A tumor is a mass of cancer cells. For instance, CNS lymphoma likely won't cause symptoms if the tumor's located in the membrane covering your brain and spinal cord. In contrast, a tumor near one or both eyes often causes vision changes. If the mass occurs near the area of your brain that controls movement, you could have weakness or coordination changes. Symptoms of CNS lymphoma may include: nause and vomiting, hearing loss, weakness in arms, legs or face, weakness affectşng one side of body, headaches, confusion, seizures. The most common reasons for admission of the patients included in our study were headache, unilateral weakness in the body and seizure. The brain parenchyma is the most common location. In our study, similar to the literature, the location of the tumors in all our patients was the brain parenchyma. Our study's findings were consistent with the literature regarding histology. However, we did not demonstrate a correlation between high-risk factors such as elevated LDH, performance scores, and double or triple expression with survival outcomes. Nevertheless, we confirmed the prognostic and predictive importance of Ki-67, in line with the literature. The lack of statistical significance for other prognostic factors may

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OP 05

RICHTER TRANSFORMATION OF CLL DISEASE: A CASE REPORT WITH CENTRAL NERVOUS SYSTEM INVOLVEMENT

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Objective: Richter transformation (Richter syndrome) is defined as the transformation of chronic lymphocytic leukaemia (CLL) into an aggressive lymphoma. Richter transformation is estimated to occur with a rate of 5-10% (1). In this article, we aimed to present a case of Richter transformation with central nervous system (CNS) involvement. **Case report:** In 2018, a 64-year-old woman who presented with lymphocytosis underwent flow cytometry examination and was diagnosed with chronic lymphocytic leukaemia (CLL). 6 cycles of Rituximab + Bendomustine treatment was administered. In 2024, the patient was admitted to the neurosurgery outpatient clinic with complaints of balance disorder, slowing of speech and facial slippage, and lesions were detected in the periventricular area at the level of basal ganglia in the brain (Figure 1). CSF examination did not reveal any findings in favour of lymphoma. A biopsy was taken from the intracranial mass. The biopsy result was reported as high-grade B- cell lymphoma. Transition from CHL to lymphoma was evaluated as Richter transformation. On PET CT examination, a 13*11 mm hyperdense lesion at the level of the left basal nucleus nucleus lentiformis, showing intense hyper metabolic activity compared to the surrounding parenchyma, showed central nervous system involvement of lymphoma. The patient was interned to the BMT ward and MATRIX protocol containing high-dose Ara-C and high-dose methotrexate with high CNS transmission was applied for a total of 3 cycles. Control brain imaging showed regression of the mass lesion observed in the previous examination. **Conclusion:** Central nervous system (CNS) infiltration is rare in Richter syndrome (RS). It has only been discussed at the case level (2). Various symptoms including headache, convulsions, diplopia, ataxia, facial paralysis and cognitive dysfunction are observed in these patients (3). In our patient, neurological findings including balance disorder, slowing of speech and facial slipping were observed. Central nervous system involvement was diagnosed by brain biopsy. When the literature is analysed, CNS involvement in these patients is very rare and usually demonstrated by CSF analysis. In a few patients, the diagnosis was made by brain biopsy as in our patient (3). In the literature, no optimal treatment has been reported in patients with Richter transformation with CNS involvement. Negative results have been shown despite treatments including high dose Methotrexate and Rituximab (1,2,3). In our case, successful results were obtained with MATRIX -C protocol including high dose Ara-C and high dose methotrexate. As a result of this treatment, regression was observed in control brain imaging. CONCLUSION: Richter transformation with CNS involvement is a rare but life-threatening condition. Close follow-up in the course of CLL, early diagnosis and initiation of appropriate chemotherapy may be life-saving. References: 1. PRONELLO, Edoardo, et al. Richter's syndrome of the central nervous system. Canadian Journal of Neurological Sciences, 2021, 48.6: 889-892. 2. Xu L, Song J Ch, Xiu H S, and et al. Richter's syndrome of the central nervous system diagnosed concurrently with chronic lymphocytic leukaemia: A case report and literature review. Medicine (Baltimore). 2018 Oct;97(41): e12701. 3. Nakanishi T, Ito T, Fujita Sh, etal. Refractory Chronic Lymphocytic Leukemia with Central Nervous System Involvement: A Case Report with Literature Review. J Blood Med. 2020; 11: 487–502.Figure 1: Intracranial mass appearance on computed tomography of the brain (marked with red colour)

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