cytotoxic agents (cyclophosphamide, cyclosporine) and rituximab (anti-CD-20 monoclonal antibody) are often used to eradicate inhibitors (7).Our patient, who presented with diffuse bleeding in the right leg and left hand, was also difficult to treat due to high factor inhibitor level and was hospitalised for approximately one month.In conclusion, AHA should be recognised and treated rapidly, as delays in diagnosis can have serious consequences. In our patient, AHA was diagnosed rapidly and treatment and bleeding control were effective.

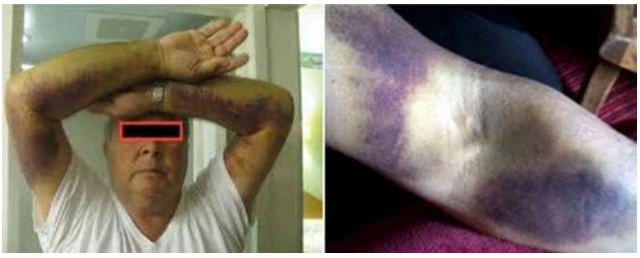

Figure 1: Ecchymosis on the forearms of both arms

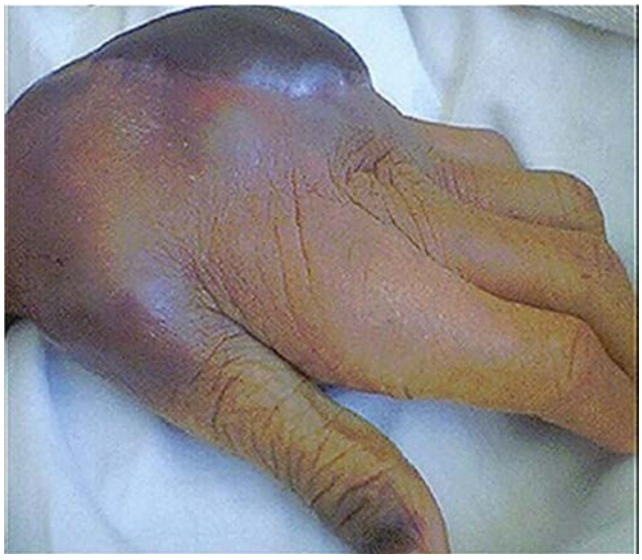

Figure 2:Ecchymosis on the dorsal left hand

https://doi.org/10.1016/j.htct.2024.11.003

OP 03

NEWLY DIAGNOSED HEREDITARY FACTOR V DEFICIENCY IN A PATIENT PRESENTING WITH DEEP VEIN THROMBOSIS: A Rare Case

Songül Beskisiz Dönen *, Vehbi Demircan, Abdullah Karakuş, Mehmet Orhan Ayyıldız

Dicle University, Faculty of Medicine, Department of Hematology

Objective: Factor V (FV) is a crucial regulator of hemostasis, functioning as both a procoagulant and an anticoagulant glycoprotein within the coagulation cascade. In plasma, FV exists as an inactive precursor, which is activated by thrombin or factor Xa into its procoagulant form, FVa, contributing to the formation of the prothrombinase complex. Additionally, FV isoforms generated via alternative splicing, notably FV-short, exhibit anticoagulant properties by interacting with tissue factor pathway inhibitor-alpha (TFPIα), thus playing a role in regulating blood coagulation. Factor V deficiency is a rare condition characterized by a bleeding tendency, with a prevalence of approximately 1:1,000,000 in the general population. Clinical presentations can range from mild mucosal bleeding to severe hemorrhagic events. However, in rare cases, thrombotic complications may also occur, highlighting the heterogeneous clinical spectrum of FV deficiency. In this case report, we will discuss a patient who presented with deep vein thrombosis (DVT) and was diagnosed with FV deficiency. **Case report:** A 43-year-old female presented to the emergency department of Dicle University Hospital on July 21, 2024, with complaints of pain and swelling in her left leg. Physical examination and lower extremity venous Doppler ultrasonography revealed an echogenic thrombus within the distal iliac, femoral, popliteal, and proximal deep crural veins, leading to a diagnosis of deep vein thrombosis (DVT). Initial coagulation tests showed prolonged prothrombin time (PT) of 246 seconds, INR of 2.21, and activated partial thromboplastin time (APTT) of 38.2 seconds, with PT being more significantly prolonged. The complete blood count was normal. The patient was admitted to the cardiovascular surgery clinic for thrombolytic therapy and underwent thrombectomy and thrombolysis. Anticoagulant therapy with bemiparin sodium (HIBOR) was initiated. Due to abnormal coagulation tests, hematology consultation was requested. Further investigations showed normal lupus anticoagulant levels (1.16). Factor assays revealed a Factor V level of <6% (normal: 70-120%), while other factor levels were within normal limits. Anticardiolipin IgM and IgG were negative, and homocysteine was normal. APC resistance was measured at 0.64 seconds (normal: 0.91-1.19). Both Factor V Leiden and Factor II mutations were homozygous normal. Antithrombin III activity, as well as Protein C and S levels, were normal. ANA and anti-dsDNA tests were negative. A repeat Factor V level confirmed it remained <6%. The patient had no history of bleeding episodes and had not experienced bleeding complications during previous childbirths or minor surgeries. This case highlights the rare identification of Factor V deficiency in a patient presenting with deep vein thrombosis, an uncommon association, illustrating the complexity and variable clinical manifestations of Factor V deficiency. **Dıscussıon:** Factor V deficiency is a rare coagulopathy, typically characterized by a bleeding tendency, with an incidence of approximately 1:1,000,000 in the general population. The disease is inherited in an autosomal recessive manner and is caused by various mutations in the FV gene. As FV plays a critical role in both procoagulant and anticoagulant pathways, its deficiency usually manifests with symptoms such as mucosal bleeding, postoperative hemorrhage, and menorrhagia. However, paradoxically, some patients with FV deficiency have been reported to develop thrombotic events. In this patient, who presented with complaints of deep vein thrombosis, the absence of a bleeding history and lack of hemorrhagic complications during past surgical procedures highlights the

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.

https://doi.org/10.1016/j.htct.2024.11.004

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

Bulut Sat [1,*], Şendağ Yaslıkaya [2], Ertuğrul Bayram [3], Berksoy Şahin [3]

[1] *Çukurova University, Clinic of Internal*
[2] *Giresun Training and Research Hospital, Clinic of Medical Oncology*
[3] *Çukurova University, Faculty of Medicine, Clinic of Medical Oncology*

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