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ASSESSMENT OF THROMBOTIC RISK AND PATHOPHYSIOLOGICAL MECHANISMS OF SPLENECTOMY-ASSOCIATED HYPERCOAGULABILITY IN PATIENTS WITH IMMUNE THROMBOCYTOPENIA

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Introduction: Immune Thrombocytopenia (ITP) is an autoimmune disease characterized by isolated thrombocytopenia in the absence of an alternative diagnosis. One of the second-line treatments after failure or resistance to corticosteroids is splenectomy. Although this therapy has high rates of durable response, there is concern about long-term complications, such as thrombosis and infection. The risk of venous thromboembolism appears to increase after splenectomy in patients with ITP; however, the reason this occurs is not well understood. The von Willebrand Factor (VWF) cycle may be altered in patients who have undergone splenectomy, as the spleen is one of the organs responsible for clearing this factor. **Objectives:** This study's primary aim is to evaluate whether patients with ITP undergoing splenectomy have an increased thrombotic risk. Subsequently, we will analyze the reasons for hypercoagulability in these patients, especially with regard to the VWF cycle. **Material and Results:** We conducted a retrospective, multicenter cohort study with patients diagnosed with primary ITP that were evaluated between 2005 and 2021. First, we compared the frequency of thrombosis in ITP in splenectomized patients to non-splenectomized patients. Subsequently, we measured factors involved in the VWF production and clearance cycle from splenectomized and non-splenectomized. We evaluated the association between splenectomy and thrombosis by Cox regression and splenectomy with Factor VIII (FVIII) and VWF antigen by linear regression. **Results:** Of the 320 patients included in the cohort, 119 underwent splenectomy, while 201 did not. The majority of the patients in both groups were female (76% in each group). The median age at diagnosis was 29 years (IQR 21–45) in the splenectomized group and 35-years (IQR 24–49) in the non-splenectomized group ($p = 0.022$). Antiphospholipid (aPL) antibody positivity occurred in 22% of the splenectomized group versus 12% of the non-splenectomized group ($p = 0.037$). The splenectomized group had a lower platelet count at diagnosis than the non-splenectomized group (17×10^9/L vs. 27×10^9/L; $p < 0.001$). The median follow-up duration was 74.2-months. The frequency of thrombosis was 12% in the splenectomized group, compared to 3% in the non-splenectomized group. Cox proportional hazard analysis revealed that splenectomy was associated with a higher risk of thrombosis (HR = 7.91; 95% CI: 2.45–25.4; $p < 0.001$). This increased risk persisted after adjusting for sex, age, aPL antibodies, and comorbidities. We also compared laboratory factors related to blood count and VWF cycle. These factors included FVIII, VWF antigen and ristocetin cofactor. We examined 47 patients (21 who had undergone splenectomy and 26 who had not). In a linear regression analysis adjusted for sex, age, and blood type, splenectomized patients demonstrated significantly higher levels of FVIII activity and VWF antigen compared to non- splenectomized patients. The absolute difference in FVIII activity was 34.7% (95% CI: 14.4–55; $p = 0.006$), and the absolute difference in VWF antigen was 43.4% (95% CI: 19.3–67.6; $p = 0.004$). **Discussion and Conclusion:** This study confirms previous results that splenectomy is associated with a higher risk of thrombosis in patients with ITP. We also showed that splenectomized patients with ITP have higher levels of VWF antigen and FVIII. This increase in VWF and FVIII activity could explain why thrombotic events occur even many years after splenectomy, possibly due to a persistent hypercoagulable state.

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DESAFIOS NO TRATAMENTO DE PÚRPURA TROMBOCITOPÊNICA TROMBÓTICA REFRATÁRIA – RELATO DE CASO

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Introdução: A Púrpura Trombocitopênica Trombótica (PTT) é uma doença hematológica caracterizada por anemia hemolítica microangiopática e trombocitopenia, podendo evoluir com disfunção orgânica. Sua fisiopatologia está associada à presença de autoanticorpos contra ADAMTS13, resultando no acúmulo de grandes multímeros do fator de von Willebrand. É considerada uma doença potencialmente fatal, com necessidade de instituição de tratamento imediato, que inclui plasmaférese, terapia imunossupressora com corticoide e Rituximabe. **Descrição do caso:** Paciente do sexo feminino, 39 anos, encaminhada para avaliação da equipe de Hematologia do Hospital das Clínicas de Ribeirão Preto para investigação de anemia e plaquetopenia. Paciente relatava quadro de cefaleia holocraniana, acompanhada de parestesia em membros superiores, dislalia e febre há 7 dias, tendo procurado atendimento médico em cidade de origem, com evidência de citopenias em hemograma. Em exames, apresentava Hb 7,1 g/dL, Ht 22%, VCM 96fl, Leucócitos 9.000 mm^3, Neutrófilos 7.200 mm^3, Plaquetas 13.000 mm^3, LDH 1519 U/L, bilirrubina indireta 1,6 mg/dL, reticulócitos 450.000 μL, coombs direto negativo, INR 0,96 e função renal sem