miocardiopatia congestiva, neuropatia periférica (incluindo a síndrome do túnel cárpico) ou autonômica, hepatomegalia ou os achados característicos de macroglossia e equimoses periorbitárias devem levar à suspeição de amiloidose AL. A coexistência de mieloma múltiplo e amiloidose AL é um fator de mau prognóstico. A abordagem terapêutica destas duas doenças exige quimioterapia e, em alguns casos, transplante medular autólogo. A utilização de novos agentes, e a rigorosa seleção de candidatos ao transplante medular melhoraram a sobrevida destes doentes atingidos por neoplasias hematológicas ainda incuráveis.

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DARATUMUMAB (DARA), CYCLOPHOSPHAMIDE, THALIDOMIDE AND DEXAMETHASONE FOR TRANSPLANT ELIGIBLE NEWLY DIAGNOSED MULTIPLE MYELOMA (TE NDMM) PATIENTS: RESPONSE RATE IMPACTS ON PFS

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Background: The four-drug combo has becoming one of the main induction treatment for TE NDMM patients (pts). We conducted a phase 2 study to assess the safety and preliminary efficacy of Cyclophosphamide (C), thalidomide (T), dexamethasone (d)-(CTd) and Dara combination. MAXDARA study has shown in the primary analysis with 21 included patients (pts), 4 and 8 pts MRD negativity after four induction cycles and two consolidation cycles post transplant, respectively. (Crusoe E et al ASH 2020, 2416 poster presentation). In the present analysis we evaluate the effect of deep response rate on PFS. **Methods:** This is a phase II, open-label single-center clinical trial. The main inclusion criteria were: TE NDMM, creatinine clearance > 30 ml/min, normal cardiac, renal and liver function and the ECOG performance status = 0 - 2. The protocol was Dara-CTd for up to four 28-day induction cycles: C-500mg oral (PO) on days 1,8 and 15, T at 100-200mg PO on days 1 to 28, (d) at 40mg PO on days 1,8,15 and 22 and Dara at 16mg/Kg/dose intravenous (IV) on days 1,8,15 and 22 during cycles 1 - 2 and every other week in cycles 3 – 4, followed by ASCT. All pts received up to four 28-day consolidation cycles that was started at D+30 after ASCT: Dara at 16mg/Kg and (d) at 40mg every other week, associated with T at 100mg PO on days 1 - 28. Dara at 16mg/Kg was used monthly as maintenance until progression or limiting toxicity. The MRD was evaluated by next-generation flow (NGF) 10−6 and PET-CT was performed when the patient obtained NGF negativity or finished consolidation. PFS outcome was estimated using Kaplan–Meier method and PFS analysis were performed based on the different response rates. (Data cut-off was April 2022) **Results:** A total of 24 pts were included. The median age being 58 (range 37– 67 years), 15 (62.5%) pts were female, 22 (92%) were non-white, 6 (25%) had an R-ISS = 1, 12 (50%) had an R-ISS = 2 and 4 (16%), an R-ISS = 3. Sixteen (66.7%) pts were IgG isotype and Six (25%) had high-risk chromosomal abnormalities [1q+, del17p, t(4;14) or t (14;16)]. To date, 23 pts have completed induction, 21 performed transplant and 14 are still in treatment after one year of dara maintenance. By ITT analysis, 22 pts (91.6%) achieved (≥ PR) after 4 induction cycles. One pts achieved SD, one MR and one sCR. Eleven (39%) pts achieved PR, and 10 (35.7%) VGPR. After two consolidation cycles, ORR was 68%, 8 (28.6%) and 11 (39.3%) pts obtained sCR and VGPR, respectively. The best response during any time of the treatment were PR in 3 (12.5%) pts, VGPR in 12 pts (50%) and sCR in 8 (33.3%) pts. In a ITT analysis, NGF MRD 10−6 negativity were observed in 4(16.6%) after four induction cycles, and in 17 (70.8%) pts after two consolidation cycles post-ASCT. In a ITT analysis, after a median follow up (FU) of 26 months, the PFS was 37months for the entire group. The median PFS comparing sCR vs ≤ VGPR were 37m vs 27m (p = 0.021), respectively, and comparing MRD negative by NGF vs no, had impacted even more on PFS with a median of 37m vs 16m (p = 0.004), respectively. The OS was not yet achieved and 83% of the patients still alive after a median FU of 27m. Four pts died from infection, two of them related with covid infection (one before transplant and one during maintenance). Another case post-transplant, considered not related to the investigational agent and one after consolidation, related to the investigational agent. **Summary/Conclusion:** The Daratumumab - CTd protocol is an active and safe regimen capable of producing deep and sustainable responses. The deeper response rate impacted on PFS, confirm that MRD negativity is critical to patient outcome.

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HEMOSTASIA E PAREDE VASCULAR: DOENÇAS DE PLAQUETAS

TROMBOCITOPENIA IMUNE: DIAGNÓSTICO E EVOLUÇÃO DE PACIENTES ACOMPANHADOS EM SERVIÇO DE REFERÊNCIA DO ESTADO DE SÃO PAULO

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Introdução: Trombocitopenia imune ou púrpura trombocitopenica idiopática (PTI), é uma doença mediada por autoanticorpo IgG causando destruição plaquetária. Acomete adultos

e crianças, no adulto a incidência é de 2-4 casos/ 100.000 pessoas/ano, pico de incidência entre 20-30 anos e prevalência maior em mulheres e após 60 anos em ambos os sexos. **Objetivo:** Traçar o perfil de adultos com PTI do Hospital do Servidor Público Estadual de SP (HSPE) e confrontar com literatura. **Materiais e métodos:** Estudo retrospectivo, análise de prontuários de portadores de PTI acompanhados regularmente. Idade, sexo, plaquetas (PLQ) ao diagnóstico, comorbidades associadas, tratamentos realizados, resposta e complicações foram avaliados, analisados e confrontados com literatura. **Resultados:** Foram avaliados 100 pacientes (78% mulheres: 22% homens), idade mediana 58 (mínimo11-84) anos, PLQ ao diagnóstico 21.500 (inserir o desvio padrão30.382,68)/mm^3. Dos pacientes analisados 22% foram mantidos com tratamento conservador, 69% submetidos a 1ªlinha com corticoterapia (CE), 7% CE+imunoglobulina (Ig) e 2% esplenectomia. Dos pacientes 1ªlinha, 56,5% tiveram resposta completa (RC): PLQ ≥100.000/mm^3, 28,9% apresentaram resposta parcial (RP): PLQ>30.000/mm^3 ou 2x maior que a contagem inicial e 14,4% sem resposta: PLQ<30.000/mm^3 ou inferior à 2x o valor inicial. Dentre os que receberam 1ª linha, associada ou não a Ig, 64,4% precisaram de 2ª linha em 30,6 meses. **Discussão:** PTI caracteriza-se pela ação imune contra glicoproteínas das superfícies plaquetária e megacariocítica. O diagnóstico é realizado com PLQ<100.000/mm^3, sem citopenia associada e exclusão de outras causas, tais como: medicações, doenças reumatologias, infecções virais, hepatopatias e causas medulares. O sintoma mais comum é o sangramento cutâneo-mucoso. Tratamento objetiva manter plaquetas adequadas para que não traga prejuízo a hemostasia primária. Intervenção terapêutica deve ser instituída quando PLQ for <20000-30000/mm^3 mesmo na ausência de sangramentos. A prevalência foi maior em mulheres (3,2:1), assim como descrito em literatura, porém com idade maior. Isto pode ser explicado pelo perfil mais idoso de usuários do HSPE. Entretanto, apesar desse perfil, nossos resultados são semelhantes aos encontrados em mais jovens. A esplenectomia demonstra cerca de 8,2% de falha a curto e 4,4% a longo prazo, nesse estudo a taxa de falha e recidiva após esplenectomia foi de 6,7 em ambas, ratificando a literatura. A transitoriedade da depleção das células B e o baixo perfil de toxicidade justificam o uso do Rituximabe no tratamento da PTI. As taxas de resposta encontrada no presente também estão em concordância com estudos anteriores que descrevem RC e RP de 43,6% e 62,5%, respectivamente. O número de pacientes utilizando análogos da trombopoetina não permitiu dados relevantes, entretanto observou-se 25% de recaída. Maior número de pacientes precisa ser analisado para conclusões definitivas, mas podemos associar à dificuldade em tomar a medição livre de períodos alimentares, bem como interação com medicamentos que diminuem sua eficácia. **Conclusão:** O manejo terapêutico da PTI é, por vezes, desafiador. Os achados encontrados foram condizentes com a literatura e reforçam que o acompanhamento regular associado às propostas terapêuticas são fundamentais para o sucesso do tratamento desta doença.

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SAFETY AND EFFICACY OF SPLENECTOMY FOR THE TREATMENT OF CHRONIC IMMUNE THROMBOCYTOPENIA

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Aims: To evaluate the short- and long-term safety and efficacy of splenectomy for the treatment of chronic immune thrombocytopenia (ITP). **Methods:** A cohort of ITP patients who underwent splenectomy between 2005 and 2020 was included and followed for up to 10 years. Descriptive analyses of complications (related to the procedure, infections, and thrombosis) and response to treatment were performed. **Results:** Out of the 87 patients included, 80.5% were women, median age at diagnosis was 33 years (IQR 24 – 50), 28.7% had hypertension and 20.7% had antiphospholipid antibodies. The median time from diagnosis to splenectomy was 19 months (IQR 6.0 – 50.0) and median follow-up duration was 61 months (27 – 108). Post-procedure complications, such as fistula, surgical site infection and bleeding requiring transfusion, occurred in 6 (7%) patients and hospital-associated thrombosis (thrombosis occurring within 90 days after splenectomy) in 3 (3.4%). Two patients had abdominal thrombosis and 1 had abdominal and deep vein thrombosis. During the follow-up, 3 patients had infections requiring hospitalization, resulting in a 5-year cumulative risk of infections of 3.4% (95% CI 0.9 – 9.1). Regarding thrombotic events after splenectomy, the 5-year cumulative incidence was 12.6% (95 CI% 6.8 – 20.9) and the median time from splenectomy to thrombosis was 20 months (IQR 1 – 42). In the subgroup analysis, patients with cardiovascular risk factors, such as hypertension, diabetes, chronic kidney disease, chronic obstructive pulmonary disease, and cerebrovascular disease, showed higher risk for developing a thrombotic event during follow-up. Sixty days after splenectomy, 72 patients (82.5%) had an overall response, of which 55 were complete (63.2%) and 17 partial response (19.3%). The median time for relapse was 12.5 months (IQR 4 - 45.3) and the 5-year treatment free survival was 55.3% (95% CI 45.8 – 66.4). At the end of the follow-up, 77 patients (88.5%) had partial or complete response and 2 (2.3%) had died. **Discussion:** This study represents a cohort of patients with ITP from a single center in Brazil that evaluated the short- and long-term safety of splenectomy, in addition to its efficacy. The recent guidelines on ITP treatment recommend splenectomy as one of the main second-line therapies following failure or relapse after corticosteroid use, along with thrombopoietin receptor agonist or rituximab. The perioperative complication rate and mortality in the presented cohort were as low as those described in previous cohort studies. The institution protocol includes pre-splenectomy vaccination but not long-term antibiotic prophylaxis in adults with ITP and, even with this approach, cumulative incidence of infection requiring hospitalization in the current cohort was approximately three times lower than that in previous cohort studies. The rate of venous thromboembolism was similar to another cohort studies and seems to be associated