<sup>2</sup> University of Health Sciences Ankara Dışkapı Yıldırım Beyazıt Training and Research Hospital, Department of Family Medicine, Ankara, TURKEY

Objective: Although plasma cell neoplasms occupy a large place in hematology practice, POEMS syndrome is very rare. Serum lambda light chain elevation and polyneuropathy, together with organomegaly, endocrinopathy, and skin lesions are the main components of the syndrome. We share our case, which we diagnosed in our clinic, with the belief that it will contribute to the literature. Case report: A 51-yearold male patient, who had no history of co-morbidity, drug use, or exposure to toxic substances, was started on supportive treatment in February 2021, who first developed the complaint of impotence. Later, he applied to the neurology outpatient clinic with complaints of weakness and weakness in the feet. After detecting polyneuropathy in his evaluation, IgG Lambda monoclonal gammopathy was detected in serum immune electrophoresis in his evaluation for etiology. Methodology: Thereupon, it started to be investigated in terms of plasma cell neoplasms. In the examinations performed, immunoglobulin levels, serum-urine kappa and lambda light chain levels, plasma increase in the bone marrow biopsies and a solitary 3.3 cm sclerotic lesion in the sacral region were detected in the PET-CT of the patient, whose ethology could not be diagnosed. Results: A tru-cut biopsy was taken from the sclerotic lesion of the patient, who was thought to be a plasmacytoma and a 20% monoclonal IgG lambda plasma increase was detected. In his physical examination, it was seen that he had increased lesions (Figure-1) and acrocyanosis (Figure-2) on the skin for the last 3-4 months. The patient's current complaints and laboratory results were evaluated with a preliminary diagnosis of POEMS syndrome (Table-1). Conclusion: POEMS syndrome is a rare disease and its exact incidence is unknown. It is frequently seen in 5-6 decades, with a median age of 51 years, and 63% of cases are male patients [1]. Chronic and excessive production of proinflammatory and other cytokines (IL-1 $\beta$ , TNF $\alpha$ , IL-6, vascular endothelial growth factor (VEGF) etc.), microangiopathy, edema, effusions, increase in vascular permeability, increase in neovascularization are important in the pathophysiology of the disease.

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## PLATELET DISEASES

PP 36

IMMUNE THROMBOCYTOPENIA RELAPSE POST COVID-19 VACCINE IN YOUNG MALE PATIENT

Hana Qasim

Hamad Medical Corporation

Case report: We report a 28-year-old Asian male patient, known for ITP and in partial remission for eighteen months, who presented to emergency department with ITP relapse (platelets count of  $1 \times 10^3$  /uL), four days after receiving the

second dose of Pfizer SARS-CoV-2 vaccine, which required treatment with intravenous immunoglobulins and dexamethasone, we discuss as well the likely underlying pathophysiology and the suggested approach in patients known for ITP who are willing to receive mRNA COVID vaccines.

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## PP 37

INTEGRATED EFFICACY RESULTS FROM THE PHASE 2 AND PHASE 3 STUDIES WITH CAPLACIZUMAB IN PATIENTS WITH ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA

Flora Peyvandi <sup>1</sup>, Spero Cataland <sup>2</sup>, Marie Scully <sup>3</sup>, Paul Coppo <sup>4</sup>, Paul Knoebl <sup>5</sup>, Johanna A. Kremer Hovinga <sup>6</sup>, Ara Metjian <sup>7</sup>, Javier de la Rubia <sup>8</sup>, Katerina Pavenski <sup>9</sup>, Jessica Minkue Mi Edou <sup>10</sup>, Filip Callewaert <sup>11</sup>, Hilde De Winter <sup>12</sup>

- <sup>1</sup> Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, and Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Milan, Italy
  <sup>2</sup> Division of Hematology, Department of Internal Medicine, The Ohio State University, Columbus, OH,
- USA

  <sup>3</sup> Department of Haematology, University College
  London Hospitals NHS Trust, London, UK

  <sup>4</sup> Department of Hematology, Reference Center for
  Thrombotic Microangiopathies (CNR-MAT), SaintAntoine University Hospital, AP-HP, Paris, France

  <sup>5</sup> Department of Medicine 1, Division of Hematology
  and Hemostasis, Medical University of Vienna,
  Austria
- <sup>6</sup> Department of Hematology and Central Hematology Laboratory, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland
   <sup>7</sup> Division of Hematology, Department of Medicine, University of Colorado—Anschutz Medical Center, Denver, CO, USA
- <sup>8</sup> Hematology Department, Internal Medicine, School of Medicine and Dentistry, Catholic University of Valencia and Hospital Doctor Peset, Valencia, Spain <sup>9</sup> Departments of Medicine and Laboratory Medicine, St. Michael's Hospital and University of Toronto, Toronto, ON, Canada
- <sup>10</sup> Clinical Development, Ablynx, a Sanofi company, Ghent, Belgium
- <sup>11</sup> Medical Affairs, Sanofi, Diegem, Belgium
- $^{12}$  Formerly Clinical Development, Ablynx, a Sanofi company, Ghent, Belgium

**Objective:** An integrated analysis based on the Phase 2 TITAN (NCT01151423) and Phase 3 HERCULES (NCT02553317) studies with caplacizumab (CPLZ) in acquired thrombotic thrombocytopenic purpura (aTTP) was performed to assess treatment

differences on efficacy and safety outcomes that may have been undetected in the individual trials. Methodology: In both trials, patients with an acute episode of aTTP were randomized to receive CPLZ or placebo (PBO) in addition to therapeutic plasma exchange (TPE) and immunosuppression. All randomized patients from both studies were included in the integrated efficacy analyses (CPLZ: n=108; PBO: n=112), and those who received at least 1 dose of the study drug were included in the safety analyses (CPLZ: n=106; PBO: n=110). Results: CPLZ significantly reduced mortality (0 vs 4 deaths; P<0.05) and refractory TTP (0 vs 8 events; P&lt;0.05) versus PBO and improved time to platelet count response (hazard ratio, 1.65; P<0.001). CPLZ also reduced the composite endpoint of TTP-related death, exacerbation, or any treatmentemergent major thromboembolic event during the treatment period (13.0% vs 47.3%; P<0.001) and median number of TPE days (5.0 vs 7.5 days) versus PBO. Mild mucocutaneous bleeding was the main safety finding for CPLZ. Conclusion: This integrated analysis provided new evidence that CPLZ prevents mortality and refractory disease in aTTP and reinforced the individual trial efficacy and safety findings. No new safety signals were identified for

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## PP 38

EPIDEMIOLOGY, TREATMENT PATTERNS, AND CLINICAL OUTCOMES AMONG PATIENTS WITH ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA (ATTP) IN THE UNITED STATES: AN ELECTRONIC HEALTH RECORDS ANALYSIS

Ayoade Adeyemi<sup>1</sup>, Filip Callewaert<sup>1</sup>, Francesca Razakariasa<sup>2</sup>, Rui de Passos Sousa<sup>1</sup>

Objective: Acquired thrombotic thrombocytopenic purpura (aTTP) is an ultra-rare, potentially life-threatening thrombotic microangiopathy (TMA). Data on epidemiology, disease management, and clinical outcomes are scarce and often heterogeneous. The aim of this study was to assess the epidemiology, disease management, and clinical outcomes in patients with aTTP in the United States. Methodology: This longitudinal retrospective observational study of the Optum-Humedica database included patients with aTTP diagnosis from October 2015 to December 2019 if they had ≥1 documented ADAMTS13 activity <10% or ≥1 aTTP episode (≥1 inpatient stay with TMA diagnosis and ≥1 therapeutic plasma exchange [TPE] during the same stay); patients with conditions that mimic aTTP were excluded. Patients were followed until loss to follow-up, end of study period, or death. All analyses were descriptive. Results: Among 666 patients with aTTP diagnosis, 302 (45%) had ≥1 aTTP episode. Annual incidence of ≥1 aTTP episode was 1.81/million (based on data from 2016 -2019). Patients with ≥1 aTTP episode received a mean of 16.7 TPE sessions; 59% used rituximab. Among patients with  $\geq 1$ 

aTTP episode, exacerbations occurred in 17% (52/302); relapse occurred in 11% (34/302). Mortality rate was 25% (167/666) among all patients with aTTP diagnosis and 14% (41/302) among patients with  $\geq 1$  aTTP episode. Conclusion: Despite treatment with TPE and immunosuppressants, the high mortality and morbidity observed in this patient population demonstrates the need for more effective therapies to improve clinical outcomes.

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## STEM CELL TRANSPLANT

PP 39

THE ROLE OF SUBPOPULATIONS OF MOBILIZED PERIPHERAL HEMATOPOIETIC STEM CELLS IN THE RESTORATION OF HEMATOPOIESIS DURING HIGH-DOSE CHEMOTHERAPY IN CANCER PATIENTS

Liudmila GRIVTSOVA<sup>1</sup>, Nikolai TUPITSYN<sup>2</sup>

<sup>1</sup> A.F. Tsyb Medical Radiological Research Center branch of the National Medical Research Radiological Center, Ministry of Health of Russia <sup>2</sup> FSBI NMITs oncology named after N.N. Blokhin "of the Ministry of Health of Russia

Objective: Mobilized peripheral hematopoietic stem cells are transplanted to cancer patients as support for high-dose chemotherapy. It is believed that the effectiveness of restoring all hematopoietic sprouts during HSC transplantation depends on the total dose of CD34+ cells. At the same time, CD34+ stem cells are a heterogeneous cell pool, including progenitor cells of different levels of differentiation and different ability to proliferate. Accordingly, it can be expected that the subpopulation composit. Methodology: We have studied of HSC subsets in 569 specimens of hemopoietic tissue (blood cells and LP cells) from 167 adult cancer patients and on 557 specimens of hemopoietic tissue from 263 pediatric cancer patients. Also, 61 samples of LP from 50 healthy HSC donors were studied. All patients were managed at bone marrow transplantation units of hematology malignancy and oncology department of N.N. Blokhin Cancer Research Center from 1996 to 2014. Results: Peripheral hemopoietic stem cells (HSC) that are transplanted to cancer patients to reduce critical pancytopenia vary in subset composition and include early polypotent precursors (CD38- and/or HLA-DR-, CD90+, CD45negative), lymphoid precursors (CD10+, CD7+, CD2+, CD19+, CD56+), megakaryocyte- (CD61+) and myeloid-committed precursors (CD117+, CD13+, CD33+). These subsets of early and committed HSC are found in different proportions in cancer patients and normal donors. Conclusion: So, the pool of mobilized HSC is heterogeneous and represented by pluripotent precursors and committed HSC in different proportions that are in variable, rather sophisticated interrelations. Mobilization effect of SC individual subsets is related with disease type. To achieve fast recovery of granulocyte lineages after HSC autologous or allogeneic transplantation one should not

<sup>&</sup>lt;sup>1</sup> Sanofi

<sup>&</sup>lt;sup>2</sup> Quinten France