lymphocytic leukemia(CLL) is a chronic B-lymphoproliferative neoplasm that is one of the most often appearances in daily hematological practice. The CD20 antigen is an attractive target in CLL as it is present on the surface of mature B-cells. Rituximab is a highly specific chimeric mouse/human anti-CD20 antibody that is widely used in the treatment of CLL and other B-lymphomas. The aim of this abstract is to describe the occurrence of Stevens-Johnson syndrome as a result of the administration of Rituximab to a patient with CLL

Case report: We report the case of a 49 years old caucasian male that four years previously was diagnosed with CLL stage A Binet. The "watch and wait" strategy was adopted at that time. But the patient disappeared from the current supervision of the hematologist and returned after 4 years with B symptoms, giant splenomegaly, hepatomegaly, peripheral lymphadenopathy, and bicytopenia (anemia and thrombocytopenia) that accompanied the lymphocytosis in peripheral blood. Stage C Binet was established, and for this patient was proposed the initiation of treatment with chemoimmunotherapy type FCR according to guidelines. But, the patient refused to administer any type of chemotherapy and in the absence of new targeted therapeutic alternatives, the most plausible solution was to initiate monotherapy with Rituximab 375 mg/m^2 weekly. On the 3rd day after the second administration of rituximab, he experienced a febrile episode 38.5 °C, fatigue, and weakness, moderate pain all over the skin, which were aggravated by a slight touch and a non-pruritic widespread maculopapular rash, which affects the oral mucosa and also the skin in the genital area, palmar and plantar region. SJS was diagnosed affecting 12% of total body surface area according to the Lund-Browder Burn calculator. Rituximab therapy was stopped and immediate treatment of SJS has started. Patients received supportive care measures including hydration, wound debridement, systemic and topical antibiotics, topical and systemic corticosteroids, nutritional support, and pain management for 4 weeks with a total recovery of skin and mucosal lesions.

Conclusion: Although SJS is a rare complication of Rituximab therapy (0.01% in a series of 167,000 patients), it remains a dreaded complication with a 30% mortality rate among patients who develop it. Recognition of clinical signs and prompt diagnosis along with complex therapy can ensure adequate recovery of the case.

https://doi.org/10.1016/j.htct.2020.09.073

CHRONIC MYELOPROLIFERATIVE DISEASES

PP 12

Concomitant essential thrombocythemia and mature B-lymphoproliferative disorder in a patient

A. Butt*, R. Qudus, N. Ali

Aga Khan University Hospital, Karachi, Pakistan

Objective: ET and B-LPD are two distinct, clonal hematologic malignancies with their concomitant existence in a single individual being exceedingly rare.

Case report: A 64-year-old male was admitted with cough, weight loss, maculopapular rash and elevated platelet counts. The rash was present on his face and trunk for 20 days and he had non-productive cough for the past two weeks. On examination, he was found to have cervical lymphadenopathy and splenomegaly (5 cm below left costal margin). His blood counts were as follows: hemoglobin 10.8 g/dl, hematocrit 37.2%, RBC mass 5.34×10^{12}/L, WBC 62.1×10^9/L, platelets 1169×10^9/L. LDH was found to be 816 IU/L and C-reactive protein was 2.43 mg/dl. Peripheral blood film showed anisocytosis, poikilocytosis, elliptical cells, tear-drop cells, nucleated red blood cells, myelocytes and metamyelocytes. Platelets were markedly increased on film. Leucoerythroblastic blood picture was noted. Suspecting a myeloproliferative disorder, additional investigations were sent while the patient was started on hydroxyurea 1gm daily and allopurinol 100 mg daily in addition to antibiotics. Bone marrow aspirate depicted increase in lymphoid cells that constituted around 35% of the total nucleated non-erythroid cell population. M:E ratio was 4:1. Bone trephine showed hypercellularity for age with overall cellularity 90 to 95%. Cellular areas exhibited increase in myeloid precursors along with prominent lymphoid cells and abundant megakaryocytes. Pan-T (CD03) and Pan-B (CD20) marker by immunohistochemistry was applied on bone trephine biopsy specimen which was interpreted as increase in B-lymphocytes. Reticulin stain showed grade MF-2 reticulin fibrosis. Overall findings were suggestive of essential thrombocythemia. In view of increased CD20 positive cells, immunophenotyping by flow cytometry was recommended. CD45 positive lymphoid cells population was 31%. This population showed reactivity to Pan-B-markers i.e. CD19 (26%), CD20 (27%), CD22 (26%), CD23 (11%) and cCD79a (30%) along with HLA-DR (12%) and CD45 (35%). Double bright positivity of CD19 and CD5 typical of CLL was absent. This population also showed positivity to lambda light chains restriction (kappa 0%, lambda 13%). Results were consistent with mature-B-lymphoproliferative disorder (B-LPD). JAK2 mutation was detected by PCR while BCR-ABL1 translocation was not detected by fluorescence in situ hybridization (FISH). Since double bright positivity for CD19 and CD5 was absent along with absence of FMC7, a diagnosis of mature-B-lymphoproliferative disorder was made. Cyclin D1 was applied on bone trephine, which was negative, and the infiltration did not reveal a follicular pattern. Ki67 was approximately 30%. A diagnosis of ET progressing to myelofibrosis and B-LPD was made. Patient was discharged in a stable condition and followed up on an outpatient basis. Ruxolitinib at a dose of 5 mg twice daily was initiated while hydroxyurea was reduced to 500 mg daily and then later to alternate day dosing. A wait and watch approach was adopted for the B-LPD Ruxolitinib was later increased to 10 mg twice daily. After a few months, ruxolitinib was switched to 15 mg daily with the counts remaining stable. The patient remains stable and asymptomatic two and half years later. The most recent blood counts show hemoglobin at 10.9 g/dl, WBC 31.1×10^9/L, and platelets 445×10^9/L.

Methodology: Retrospective review of case.

Conclusion: We report a rare case of ET with concomitant B-LPD. The patient is stable on Ruxolitinib and is on wait and watch approach for B-LPD.

https://doi.org/10.1016/j.htct.2020.09.074

PP 13

Acute phase reactants in chronic inflammation leading to secondary myelofibrosis in polycythemia vera and essential thrombocytosis

E. Aladag[1], I. Haznedaroglu[1], N. Sayinalp[1], H. Demiroglu[1], H. Goker[1,*], S. Aksu[1], O. Ozcebe[1], A. Ayhan[2], Y. Buyukasik[1]

[1] *Hacettepe University Department of Hematology, Ankara, Turkey*
[2] *Hacettepe University Department of Pathology, Ankara, Turkey*

Objective: Polycythemia vera and essential thrombocytosis are chronic and progressive myeloproliferative neoplasms characterized by a clonal increase in hematopoietic stem cells in the bone marrow. Myelofibrosis in the bone marrow has been shown to be secondary to an inflammatory process.

Methodology: To investigate the association between the secondary myelofibrosis and acute phase reactants in patients with polycythemia vera and essential thrombocytosis. Fourty-six PV and 28 ET patients without myelofibrosis above Grade 1 were included in the present study. Bone marrow evaluations were performed retrospectively. C-reactive protein, ferritin, and albumin levels were measured.

Results: C-reactive protein (0.55 ng/L vs. 4.2 ng/L, $p < 0.001$) and ferritin (18.5 ng/mL vs. 118 ng/mL, $p = 0.001$) levels in patients with secondary myelofibrosis were found to be increased compared to baseline levels. Mean albumin levels in patients with secondary myelofibrosis, and CRP, ferritin, and albumin levels in patients without secondary myelofibrosis were similar at the diagnosis and at last visit. There were also similar the baseline levels of CRP, ferritin, and albumin between the patients with and without secondary myelofibrosis.

Conclusion: The increase in CRP and ferritin, which are indicators of chronic inflammation, may be used to show the inflammation and relevant secondary fibrosis in the bone marrow. Due to the similar CRP, ferritin, and albumin levels at the diagnosis, the prediction for the development of the secondary myelofibrosis is not possible in the present study.

https://doi.org/10.1016/j.htct.2020.09.075

PP 14

Polycythemia vera: updates in diagnosis and treatment outcomes

L. Musteata[1,*], S. Pinzari[2], V. Musteata[1], N. Sghibneva-Bobeico[2], A. Dorogan[2]

[1] *State University of Medicine and Pharmacy, Chișinău, Republic of Moldova*
[2] *Institute of Oncology, Iași, Romania*

Objective: The objective of the study was to analyze the contemporary clinical and laboratory features of polycythemia vera (PV), as well as to evaluate the short- and long-term results of different treatment options.

Methodology: The clinico-hematological evolution features, complications, short- and long-term results of cytoreductive treatment were evaluated in a group of 114 PV patients, aged at 28–78 years old, who were followed up at the Institute of Oncology of Moldova between 1987–2019. The diagnosis was proved by the bone marrow biopsy and quantitative detection of JAK2 V617F mutation in pending cases. Physical and histopathologic examinations were associated with the repeated complete blood counts and abdominal ultrasound scan. The treatment included phlebotomies and cytoreductive chemotherapy with busulfan (56 patients) and hydroxycarbamide (58 patients) in standard doses. The life-table method was used for Kaplan–Meier Survival Analysis in order to evaluate the long-term results of treatment.

Results: The disease was commonly diagnosed in males – 66 (57.9%) patients. The females prevailed in the age groups of 40–49 years (31.3% versus 24.6% in males) and 60–69 years (25% versus 19.8% in males). The disease span from the onset of the initial clinical manifestations until the diagnosis lasted 4–9 months (median – 5.8 months) in the majority of patients (86.8%), that led to the development of thromboembolic complications in 28.1% of cases. The diagnosis was proved in stage IIA disease in 105 (92.1%) patients, IIB in 9 (7.9%) patients. The skin hiperemia was registered in 112 (98.3%) cases, scleral congestion – in 109 (95.6%), splenomegaly – in 77 (67.5%), erythromelalgia – in 71 (62.2%), aquagenic skin itching – in 68 (59.6%), hepatomegaly – in 61 (53.5%), vascular thrombosis – in 32 (28.1%). The complete blood count revealed the increase of hemoglobin (18.0–23.5 g/dL) and red cells (5.5–6.7 × 1,000,000 [MICRO]/L). The platelets range was 180–1690 × 1000 [MICRO]/L, leukocytes range – 5.1–21.3 × 1000 [MICRO]/L. Leukocytosis occurred in 69 (60.5%) patients, thrombcytosis – in 61 (53.5%). The bone marrow biopsy detected a hyperplasia due to the proliferation of erythroid, granulocyte and megakaryocyte cell lines. The study of short-term results asserted the complete remissions in all cases under chemotherapy combined with phlebotomies. The overall one-, 5-, 10- and 15 year was 100%, 98.6%, 85.9% and 67.1%, respectively. 73 (64.04%) patients remain in stage II disease after the treatment during 5–26 years of follow-up. The survival median was not reached.

Conclusion: The reluctant evolution, progressive growth of hemoglobin and red cell count, gradual increase of blood hyperviscosity and the lack of hemato-oncological vigilance of primary care physicians may lead to the development of