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631.MYELOPROLIFERATIVE SYNDROMES AND CHRONIC MYELOID LEUKEMIA: BASIC AND TRANSLATIONAL

Co-Occurrence of Familial Mediterranean Fever and Hematologic Malignancies: Case Report Series

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Introduction: Familial Mediterranean Fever (FMF) is an autosomal recessive genetic disorder primarily found among individuals of Mediterranean descent, including Armenians, Italians, Greeks, Arabs, Turks and Jews. In the Armenian population, approximately 90% of FMF cases are associated with three main mutations (M694V, V726A, and M680I) in two alleles of MEFV gene, while more than 300 MEFV variants have been reported worldwide. The homozygous M694V genotype is associated with frequent renal failure due to amyloidosis. Clinical manifestations of FMF include periodic attacks of diffuse serositis with pain and fever that are treated with colchicine or biologic therapy. There is a scarcity of data on patients with FMF and hematologic malignancies. Limited data makes it difficult to generate evidence-based recommendations for the management of these patients, which is mostly based on expert opinions or few case reports. Here we report the case series of patients from Armenia with simultaneous presence of FMF and hematologic malignancies (FMFHM) and evaluate their management challenges.

Methods: Data was collected from the Armenian National Registry of Blood Disorders, at the Hematology Center after Prof. R. Yeolyan, and through personal interviews conducted with each patient. Hematology Center is the only institution in Armenia managing hematologic malignancies. We have analyzed 8 patients diagnosed with both FMFHM during 2000-2020, treated in the pediatric (1 patient) and adult (7 patients) hematology departments.

Results: 8 patients with FMFHM were identified, from which 4 were diagnosed with myeloproliferative neoplasms (MPN), 2 with acute leukemia (AL), and 2 with non-Hodgkin lymphoma (NHL). The median age of patients was 46, range 20-60 years. Only 5 (62.5%) of patients were regularly taking colchicine for FMF, while the rest refused it. Four MPN patients received treatment only with oral medications (tyrosine kinase inhibitors, hydroxyurea). Autologous hemopoietic stem cell transplantation (auto-HSCT) was performed in 1 NHL patient after indicated 1 st and 2 nd line immuno-chemotherapy. Another 2 AL patients and one NHL patient received protocol-based respective chemotherapy regimens.

Moderate gastrointestinal (GI) complications (e.g., epigastric pain, nausea, vomiting, intestinal discomfort) was experienced by 7 (87.5%) patients. Allergic reactions (e.g., moderate skin rash, itching) were seen in 5 patient (62.5%), of which one developed severe anaphylactic shock. A patient with FMF and PMF had ischemic stroke caused by thrombi. Arterial hypertension occurred in 3 (37.5%). Only in one patient reduction of colchicine dosage was performed (1mg/day was reduced to 0.6mg/d), because of interaction with imatinib. The rest of patients received chemotherapy and colchicine treatment as indicated. All 8 patients are alive, 5 of them are in hematologic or molecular remission, two of which received concurrent colchicine treatment. One patient in remission developed renal failure due to FMF (colchicine was refused). Two patients with MPN treated with colchicine experienced disease stabilization.

Conclusion: In conclusion, the concurrent appearance of FMFHM may potentially increase the risk of GI side effects, allergic complications, and arterial hypertension, however definitive conclusions require further investigations in larger patient cohorts. While the overall treatment responses were largely positive, the association between MEFV and HM mutations warrants

deeper exploration to provide more precise drug administration recommendations for colchicine and cancer medications. Future studies in this area will contribute to a better understanding of the interplay between these conditions and guide optimal treatment strategies.

Disclosures Kazandjian: *Plexus Communications:* Ended employment in the past 24 months, Honoraria; *MJH Life Sciences:* Current Employment, Honoraria; *MMRF:* Ended employment in the past 24 months, Honoraria; *Bridger Consulting Group:* Consultancy, Honoraria; *Curio Science:* Ended employment in the past 24 months, Honoraria; *Bristol Myer Squibb:* Consultancy, Honoraria; *Karyopharm Therapeutics:* Current Employment, Speakers Bureau; *Sanofi:* Consultancy, Honoraria; *Aptitude Health:* Consultancy, Honoraria; *Arcellx:* Consultancy, Current Employment, Honoraria; *Aperture Medical Technology, LLC:* Consultancy, Honoraria; *Alphasights:* Consultancy, Honoraria.

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