Challenges of the management of Primary Myelofibrosis in Armenia

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Background

Myelofibrosis (MF) is a myeloproliferative neoplasm (MPN) characterized by stem cell-derived clonal myeloproliferation that is often but not always accompanied by *JAK2*, *CALR*, or *MPL* mutations.

Objective

The aim of our work was to evaluate the diagnostic and therapeutic challenges and estimate the survival for the patients with primary myelofibrosis (PMF) in Armenia for the last decade.

Materials & Methods

The data was collected from the medical records of all patients diagnosed with PMF in Armenia from 01. January 2010 to 31. December 2020. Survival analysis was performed using Kaplan- Meier method.

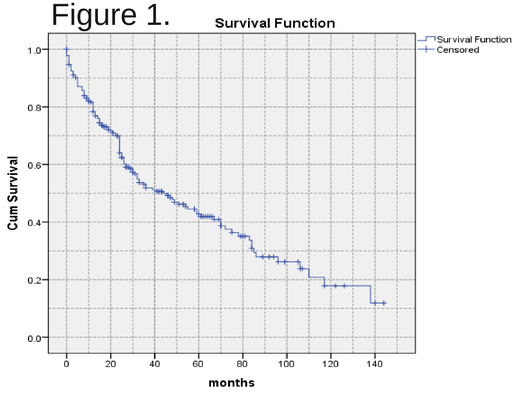
Results

The total number of patients was 238 of which 112 (47.1%) patients were alive at the time of data collection. The annual incidence was 0.95 case per 100.000. Median age was 60 years (28-88). 42.9% (102) of patients were female. In all 112 patients the diagnosis was based on the typical megakaryocyte changes accompanied by ≥ grade 2 reticulin/collagen fibrosis of bone marrow.

JAK2, CALR, MPL mutations, or presence of other clonal markers was estimated only in limited number of patients: JAK2 in 10.7% (11) patients of which 6 were positive; MPL in one patient, which was positive; CALR and other clonal markers were not estimated in any patient. BCR/ABL was estimated in 12 patients, all were negative.74.1% (83) patients received treatment with hydroxycarbamide; 5.3% (n=6) ruxolitinib; 6.3 % (n=7) interferon alfa-2; 8% (n=9) were under observation; 6.3% (n=7) unknown treatment. None of patients underwent allogeneic hematopoietic stem cell transplantation (HSCT), which is not available in Armenia. Diagnostic and treatment limitations are due to financial reasons, majority of expenses are out of pocket.

Median survival of patients was 44 months (95%CI: 30-58 months), 5-year overall survival was 40 % (figure 1).

Figure 1.



Conclusion

This is the first comprehensive analysis of PMF in Armenia. It showed significant diagnostic and therapeutic limitations which led to inferior outcomes.