P1039 PALPABLE SPLEEN SIZE IS PROGNOSTIC IN PRIMARY BUT NOT SECONDARY MYELOFIBROSIS

Topic: 16. Myeloproliferative neoplasms - Clinical

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Background: Splenomegaly is a clinical hallmark of primary (PMF) and secondary myelofibrosis (SMF). Although not a part of disease specific risk scores, spleen size represents disease burden and correlates with unfavorable disease features. Spleen size reduction is a function of baseline spleen size and was recently shown to be independent prognostic factor in composite PMF and SMF cohort of ruxolitinib treated patients.

Aims: To estimate prognostic properties of palpable splenomegaly and potential differences between PMF and SMF patients.

Methods: We analyzed a multicentric cohort of 191 patients from 6 Croatian centers with either PMF or post polycythemia vera (PV) and essential thrombocythemia (ET) SMF diagnosed in period from 2004-2021 who had available data on palpable spleen size. Diagnoses were established according to 2016 and 2008 criteria. Risk was stratified according to the DIPSS in PMF and according to the MySEC-PM in SMF patients. Palpable spleen size was assessed at the time of diagnosis or referral and stratified in three categories: non-palpable, <10 cm and ≥10 cm.

Results: A total of 191 patients were analyzed, among them 144 PMF (63 early, 81 overt fibrotic) and 47 SMF (27 post PV; 20 post ET). Median age was 67 years, there were 64% males. Intermediate-2 or high risk disease was present in 45% PMF and 49% SMF patients. Non-palpable spleen, <10 cm and ≥10 cm splenomegaly were present in 31%, 43% and 26% PMF patients, respectively, and 23%, 49% and 28% SMF patients, respectively. Palpable spleen size was similar between patients with PMF and SMF (median 3 vs 4 cm, P=0.325). Patients with palpable spleen ≥10 cm were significantly grouped among patients with higher risk disease in PMF (P=0.026) but not SMF (P=0.654). Degree of bone marrow fibrosis was significantly associated with higher spleen size in PMF (P=0.008) but not SMF (0.809). On the contrary, JAK2 mutational status had no significant association with spleen size in PMF (P=0.847) but was significantly associated with higher spleen size in SMF (P=0.035).

Regarding overall survival, larger spleen size was significantly associated with shorter overall survival in PMF (P<0.001) but not SMF (P=0.825) as shown in Figure. Time to thrombosis had no significant association with larger spleen size in neither PMF nor SMF (P>0.05). Time to bleeding was significantly shorter in PMF patients with both <10 cm and ≥10 cm in comparison to non-palpable spleen (P<0.05) whereas palpable spleen size had no significant association with in SMF patients (P>0.05).

Image:
Summary/Conclusion: Various clinical features are differently associated with palpable spleen size in PMF and SMF patients. Larger spleen size is prognostic of worse survival and higher bleeding risk in PMF but not SMF patients. This might have repercussions on prognostic scores based on palpable spleen size that might differently perform in PMF and SMF patients.