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LETTERS

Is life expectancy of polycythemia vera patients clearly different from that of the general population?

To the Editor:

In a recent issue of the *American Journal of Medicine*, Passamonti et al reported a significantly higher mortality for polycythemia vera (PV) patients than in a sex-, age-, and calendar year-matched Italian general population in a retrospective analysis of 396 PV patients, using standardized mortality ratio (SMR).[1] Thrombosis was the most frequent complication and the main cause of death, and a history of thrombosis was the only adverse prognostic factor for survival. By contrast, survival of patients with essential thrombocythemia was similar to that of the general population.

We previously published the final analysis of a prospective study that enrolled 179 PV patients with the longest mean follow-up reported to date of 11.4 years.[2] In our study, SMR for the entire cohort, as well as for the different age and sex subgroups, were not statistically different from those of the French population. We also found that hyperleukocytosis was a significant risk factor for evolution to leukemia and shortened survival. A trend for such unfavorable impact of hyper leukocytosis on leukemic evolution ($P = 0.065$) was also reported in the recent analysis of the European Collaboration on Low-dose Aspirin in Polycythemia Vera (ECLAP) prospective study.[3]

Reaching a balance between the vascular risk and the possible leukemogenicity of cytoreductive treatments is a major aim in the therapeutic strategy of PV. Evaluation of life expectancy of PV patients, identification of prognostic factors, and precise determination of very long-term complications are crucial for the management of this long-lasting disease. In our study, the incidence of thrombosis (15% at 10 years) was even lower than observed in the French matched-control population, showing that a careful management of PV might reduce the vascular risk associated with the disease. All our patients had been homogeneously treated with a cytoreductive agent (pipobroman) with a very high response rate of 98%. In the Passamonti et al study, various therapeutic strategies were used and the response rate was not documented, making difficult the interpretation of the high incidence of thrombosis reported. Furthermore, the somewhat shorter follow-up (9.6 years versus 11.4 years in our study), the smaller number of deaths observed (33% compared to 54% in our study), and the retrospective nature of their study could have biased the data obtained.

Finally, controversies concerning outcome of patients with myeloproliferative disorders stress the need for long-term data obtained from prospective studies in chronic malignancies that cannot be cured with currently available drugs.

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References

1. Passamonti F, Rumi E, Pungolino E, et al. Life expectancy and prognostic factors for survival in patients with polycythemia vera and essential thrombocythemia. *Am J Med.* 2004;117:755–761.
2. Kiladjian JJ, Gardin C, Renoux M, Bruno F, Bernard JF. Long-term outcomes of polycythemia vera patients treated with pipobroman as initial therapy. *Hematol J.* 2003;4:198–207.
3. Finazzi G, Caruso V, Marchioli R, et al. Acute leukemia in polycythemia vera. An analysis of 1638 patients enrolled in a prospective observational study. *Blood.* 2005;105:2664-2670.

The Reply:

Kiladjian et al state that the results of their study showed a standardized mortality ratio (SMR) of 1.0 and a 15-year risk of thrombosis of 21.8% in 164 polycythemic patients.[1] In our cohort of 396 patients with polycythemia vera, SMR was 1.6 and the 15-year risk of thrombosis was 27%.[2]

The patient cohort included 130 patients with polycythemia vera and 34 individuals with idiopathic erythrocytosis. Idiopathic erythrocytosis represents a grey zone between diagnosis of polycythemia vera and that of secondary erythrocytosis.[3] The fact that 20% of patients in the Kiladjian study were classified as idiopathic erythrocytosis might explain the lower SMR and the slightly lower risk of thrombosis compared with our study that included only patients with polycythemia vera.

To estimate SMR, Kiladjian et al compared the observed mortality with the probability of death in an age- and sex-