骨髓增生腫瘤之臨床處置

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Thromboembolic events were the most frequent complications in patients with polycythemia vera (PV) and essential thrombocythemia (ET). In prospective studies, the rate of thrombosis ranged between 2-4% per patient-year in ET patients, whereas cardiovascular mortality accounted for 1.5 deaths per 100 patients per year in PV. Therefore, prevention of thromboembolism is the major goal in our management of EV and PV patients.

Traditionally, risk categorization for patients with ET and PV is based on the presence or absence of the following two factors: advanced age (60 years or older) and history of thrombosis. Patients with either one of the 2 factors are considered high-risk. Controlled studies have confirmed the anti-thrombotic value of low-dose aspirin in PV, and such therapy has also been shown to be effective in alleviating vasomotor (microvascular) disturbances associated with ET. A recent study successfully demonstrated the benefits of a target hematocrit of <45% in PV patients. Therefore, the use of once-daily aspirin in low-risk ET and PV patients and regular phlebotomy with the hematocrit target of <45% in PV patients are justified. On the other hand, additional therapy with cytoreductive agents (such as hydroxyurea) in high-risk patients can further reduce the incidence of vascular events. Special attention should be paid when considering the application of hydroxyurea in patients below the age of 40 because of the uncertainty of the leukemogenetic potential of this drug in this population. Because patients with extreme thrombocytosis (> 1,000 x 10^9/L) carried an extraordinarily high risk of bleeding, use of once-daily aspirin in these patients should be very careful and is best preceded by cytoreductive therapy.

In spite of the aforementioned recommendations, some controversies do exist. For examples, some consider *JAK2*V617F mutation per se a risk factor for thrombosis. Do we manage patients with or without the mutation differently? The newly identified somatic mutation of *calreticulin* (*CALR*) is associated with less thromboembolic events and a longer thrombosis-free survival in patients with ET. Are we over-treating these patients with cytoreductive agents and/or once-daily aspirin? And finally, the risk/effectiveness of the use of cytoreductive agents has never been tested in low-risk patients. Future randomized controlled trials carefully addressing these issues are urgently needed to solve the uncertainties surrounding our daily practice.