Abstract

Hemophagocytic lymphohistiocytosis (HLH; hemophagocytic syndrome) is a rare syndrome of potentially fatal, uncontrolled hyperinflammation. Allogeneic hematopoietic stem cell transplantation (allo-HSCT) is indicated in primary, recurrent or progressive HLH, but information about its outcomes in the adult population is limited. We obtained data about 87 adult (≥18 years of age) patients retrospectively reported to the EBMT. The median survival time was 13.9 months. The three and five-year overall survival (OS) was 44% (95% CI 33–54%). Among 39 patients with a follow-up longer than 15 months, only three died. Relapse rate was 21% (95% CI 13–30%), while NRM reached 36% (95% CI 25–46%). Younger patients (<30 years of age) had better prognosis, with an OS of 59% (95% CI 45–73%) at three and five years vs 23% (95% CI 8–37%) for older ones. No difference in survival between reduced and myeloablative conditioning was found. To our knowledge, this is the largest report of adult HLH patients who underwent allo-HSCT. Patients who survive the first period after this procedure can expect a long disease-free survival. Both reduced intensity and myeloablative conditioning have therapeutic potential in adult HLH.