Survival Rate of Adult Patients with Hemophagocytic Syndrome at Siriraj Hospital

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**Background:** Hemophagocytic syndrome or hemophagocytic lymphohistiocytosis (HLH) is an uncommon, life-threatening condition in adults and delay in diagnosis and treatment is associated with high mortality. Data regarding survival rate of adult patients with HLH in Thailand are limited.

**Objective:** To study survival rate of hemophagocytic syndrome at Siriraj Hospital.

**Methods:** A retrospective chart review of 54 patients aged > 15 years diagnosed of HLH at Siriraj Hospital between 2006 and 2016 was performed. Data including baseline demographics, clinical manifestation, laboratory investigation, HLH diagnostic criteria, cause of HLH, treatment, and clinical outcome were assessed. Primary outcome was survival rate of HLH. Secondary outcome was the factor associated with death.

**Results:** In total, 33 out of 54 patients were correctly diagnosed HLH by using HLH-2004 criteria (≥ 5 criteria). The median age was 37.9 years (range 16-80). There were 19 women and 14 men. The known causes of HLH included lymphoma (67.6%) and infection (33.3%). The survival rate was 57.6%. In univariate analysis, the significant predictors for death were lymphoma (P=0.024), D-dimer level <6000 ng/mL (P=0.005) and PT ≥15 sec (P=0.017). In a multivariate analysis, the significant predictor for death was D-dimer level <6000 ng/mL (P=0.007; OR 35, 95\%CI, 2.6-465.4).

**Conclusion:** Survival rate of HLH at Siriraj Hospital is 57.6\%. The factor associated with poor outcome is D-dimer level <6000 ng/mL.

**Keywords:** Hemophagocytic syndrome, HLH