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abstract

Outcome and Prognostic Factors of Hemophagocytic Lymphohistiocytosis in Children: Experience From a Low- and Middle-Income Country

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Introduction: HLH presents as a febrile illness with multi-organ involvement. Initial signs and symptoms of HLH can mimic common infections, fever of unknown origin, hepatitis, or encephalitis, making the estimation of real incidence difficult worldwide. In children, mostly inherited immune deficiencies lead to this syndrome. Some reports estimate 1.2 cases per million children per year with infants being most susceptible. Therefore, a high degree of suspicion must be exercised for early diagnosis, aggressive treatment, and supportive care for HLH. Hemophagocytic lymphohistiocytosis (HLH) is a life-threatening condition especially in low- and middle income countries (LMICs). This study was done to evaluate the outcome and prognostic factors of HLH in patients presenting to a tertiary care hospital in a resource limited country.

Methodology: The study was carried out at the Paediatric Oncology Department of Combined Military Hospital (CMH) in Rawalpindi, Pakistan. All cases of HLH, from one month to 15 years of age enrolled between January 1, 2013 to June 30, 2023, were included.

IBM SPSS Statistics for Windows, version 25.0 (released 2017, IBM Corp., Armonk, NY) was used for statistical analysis, and t-test and chi-square tests were used for comparison between continuous and categorical variables. Frequencies and percentages were calculated for categorical variables.

Results: Out of 115 patients, seven (6%) abandoned the treatment. The data of 108 cases, including 58 males (53.7%), were analyzed. The mean age at diagnosis was 31.5 ± 39.03 months. The mean time to reach a pediatric oncologist was 30.20 ± 22.15 days. Fever and pallor were common symptoms occurring in 107 (99.1%) and 98 (90.7%) cases, respectively. Jaundice was present in 44 (40.7%), visceromegaly in 64 (59.3%), and bruising/bleeding in 16 cases (14.8%). Twenty-six (24.1%) patients underwent hematopoietic stem cell transplant (HSCT), out of which 17 (65.4%) children were cured. Overall survival at two years, five years, and 10 years was 38%, 37%, and 36.1%, respectively. Disease-free survival at two years, five years, and 10 years was 33.3%, 32.4%, and 31.5%, respectively.

Conclusion: HLH leads to high mortality due to delayed or misdiagnosis in LMICs. Early diagnosis and early referral to a pediatric oncologist is the detrimental factor in survival for HLH. HSCT is the treatment of choice for primary, refractory, or relapse cases.