Malignancy-associated haemophagocytic lymphohistiocytosis in children and adolescents.

Abstract:
Haemophagocytic lymphohistiocytosis (HLH) in the context of malignancy is mainly considered a challenge of adult haematology. While this association is also observed in children, little is known regarding inciting factors, appropriate treatment and prognosis. We retrospectively analysed 29 paediatric and adolescent patients for presenting features, type of neoplasm or preceding chemotherapy, treatment and outcome. Haemophagocytic lymphohistiocytosis was considered triggered by the malignancy (M-HLH) in 21 patients, most of whom had T- (n = 12) or B-cell neoplasms (n = 7), with Epstein-Barr virus as a co-trigger in five patients. In eight patients, HLH occurred during chemotherapy (Ch-HLH) for malignancy, mainly acute leukaemias (n = 7); an infectious trigger was found in seven. In M- and Ch-HLH, median overall survival was 1.2 and 0.9 years, and the 6 month survival rates were 67% and 63%, respectively. Seven of 11 deceased M-HLH patients exhibited active malignancy and HLH at the time of death, while only two out of five deceased Ch-HLH patients had evidence of active HLH. To overcome HLH, malignancy- and HLH-directed treatments were administered in the M-HLH cohort; however, it was not
possible to determine superiority of one approach over the other. For Ch-HLH, treatment ranged from postponement of chemotherapy to the use of etoposide-containing regimens.