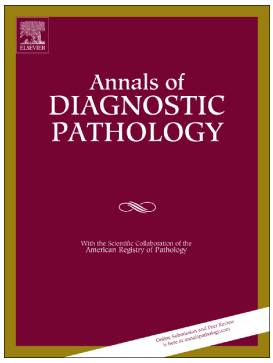

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Lymphoid Malignancy-Associated Hemophagocytic Lymphohistiocytosis: Search for the Hidden Source

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Abstract

Secondary hemophagocytic lymphohistiocytosis (HLH) is an uncommon, but life-threatening syndrome of highly stimulated and ineffective immune dysregulation. It is not a disease entity by itself and the current diagnosis of secondary (acquired) HLH is based on constellation of nonspecific clinical and laboratory parameters indicative of overactive immune response. The presenting symptoms are often nonspecific and could potentially be missed, leading to a fatal outcome. Patients with malignancy-associated HLH have a relatively unfavorable overall survival compared with non-malignancy-associated HLH. In this retrospective study, nine adult patients with secondary HLH were identified. Of these four cases were associated with a malignancy and despite a high degree of suspicion; the