Ten Best Readings Relating to Rare Lymphoproliferative and Histiocytic Diseases


Single-agent chemotherapy is typically effective in HIV-associated multicentric Castleman disease; however, chemotherapy cannot be discontinued in most patients. Rituximab has been shown to be effective and safe in patients with HIV infection and chemotherapy-dependent, multicentric Castleman disease.


Siltuximab in combination with best supportive care was superior to best supportive care alone for patients with symptomatic multicentric Castleman disease and was well tolerated with prolonged exposure.


Researchers discovered that treatment with SL-401 consisting of the catalytic and translocation domains of diphtheria toxin fused to interleukin 3 resulted in a high response rate among patients with blastic plasmacytoid dendritic cell neoplasm.


These data demonstrate that most patients with localized disease are treated similar to soft-tissue sarcoma with primary surgical resection with or without radiation. No chemotherapy data were available in the Surveillance, Epidemiology, and End Results database. The roles of chemotherapy and radiation therapy remain unclear.


In this large series of adults with secondary hemophagocytic lymphohistiocytosis (HLH) treated at a single tertiary care center, patients with low levels of serum albumin and tumor-associated HLH had poor survival rates. HLH remains elusive and challenging to health care professionals who must maintain a high index of suspicion. The recent discovery of several novel diagnostic and therapeutic modalities may improve outcomes of adult patients with HLH.


Molecular studies using paraffin-embedded archival samples showed no evidence of a positive association between Kikuchi–Fujimoto disease and viral infections, including Epstein–Barr virus and human herpesviruses 6 and 8.


High prevalence, recurrent BRAF mutations in Langerhans cell histiocytosis indicate that it is a neoplastic disease that may respond to RAF pathway inhibitors.


Reactivations of multisystem Langerhans cell histiocytosis (MS-LCH) are reduced by prolonging initial chemotherapy. In addition, the previously high mortality rates of children at high risk for MS-LCH have been reduced.


The aim of this study was to assess the results of hematopoietic stem cell transplantation (HSCT) in refractory Langerhans cell histiocytosis. The researchers concluded that HSCT for refractory Langerhans cell histiocytosis can be highly toxic but can also achieve sustained disease control.


In this review, the authors discuss recent progress in the use of hematopoietic stem cell transplantation in patients with hemophagocytic lymphohistiocytosis and potential future strategies, including the use of reduced intensity conditioning regimens.