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1711. Histoplasmosis-Associated Hemophagocytic Lymphohistiocytosis: A Case Series and Review of the Literature
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Background. Histoplasmosis is an endemic fungal disease with a spectrum of presentations from asymptomatic to disseminated infections. Histoplasmosis-associated hemophagocytic lymphohistiocytosis (HLH) is a rare disorder with limited data regarding treatment and outcome. We described the clinical features, treatment, and outcomes of five patients. This review also summarized the current literature about presentation, treatment, and outcome of this infection-related HLH entity.

Methods. We searched the electronic medical records for patients with histoplasmosis-associated HLH at our institution from January 1, 2006 to September 30, 2017. Diagnosis of HLH was confirmed and by chart review according to HLH-04 criteria. We also searched the current literature for case reports and case series of this entity.

Results. We reported five cases of histoplasmosis-associated HLH during this period. All patients were diagnosed after 2010, and this may be explained in part by increased awareness of this entity. The literature review yielded 60 cases of histoplasmosis-associated HLH. Among all patients (65 patients), the most common underlying condition was HIV in 61% of all patients. The majority of histoplasmosis patients were treated with amphotericin B formulation in 81%. The specific treatment for HLH was as follows: nine patients received steroids only; six patients received intravenous immunoglobulin (IVIG) only; three patients received dexamethasone and etoposide; two patients received etoposide, dexamethasone, and cyclosporine; two patients received steroids and IVIG, and one patient received Anakinra and IVIG. The mean mortality rate was 31% with most of the deaths occurring within 2 weeks of hospital admission.

Conclusion. Histoplasmosis-associated HLH among adults is an uncommon but aggressive disease with multiorgan involvement. Early antifungal therapy with a lipid formulation amphotericin B is the most important part of the management. Initial HLH-specific immunosuppressive therapy with regimens such as the HLH-94 protocol is usually individualized.