B46

Meenakshi Kurup M.D. Advisor(s): Neda Valizadeh M.D.

Co-author(s): Elina Shtridler, Aye Mon Thida

Hemophagocytic Lymphohistiocytosis in an HIV-Negative Adult with Penicillium Non-Marneffei Infection

Background: Hemophagocytic Lymphohistiocytosis (HLH) is a life-threatening syndrome characterized by excessive immune activation. Diagnosing HLH is challenging due to its variable presentation and crucial due to the high mortality rate. This report presents a unique case of HLH secondary to Penicillium non-marneffei in an HIV-negative patient.

Case Presentation: An 85-year-old Haitian male presented with presyncope and fever. Extensive workup ruled out infectious, rheumatological, and malignancy causes. His fever persisted despite antibiotics. Computed tomography of the chest and abdomen (CT) showed mediastinal lymphadenopathy with varying stages of calcifications. An erythematous, nontender, nonpruritic nodule developed on his right arm, which spread to his extremities. A punch biopsy showed non-necrotizing granulomatous changes, without evidence of microorganisms. As such he was diagnosed with Sarcoidosis and treated with steroids for defervescence and transaminitis and was discharged. Three days post-discharge, he was readmitted for Enterococcus faecalis bacteremia. Recurrent fevers, with worsening transaminitis, elevated inflammatory markers, and cytopenia, persisted and deteriorated despite the resolution of bacteremia. His calculated H-score was 157. A 5-day course of IVIG 2g/kg was initiated. He continued to deteriorate, with multiorgan dysfunction and a rising H-score (233, corresponding to a 98-99% likelihood of HLH). Soluble interleukin-2 receptor was elevated at 11609 U/mL. A prolonged incubation of fungal cultures from the previous punch biopsy grew Talaromyces species non-marneffei. Amphotericin B, Voriconazole, and high-dose dexamethasone were initiated. Etoposide and Rituximab were withheld due to infectious etiology. Amphotericin-B was stopped after one dose due to hepatotoxicity. Despite treatments, he deteriorated and expired.

Conclusion: HLH demands early recognition and prompt treatment to prevent significant mortality.