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623.MANTLE CELL, FOLLICULAR, AND OTHER INDOLENT B CELL LYMPHOMAS: CLINICAL AND EPIDEMIOLOGICAL

Description of Patients with HIV and Multicentric Castleman's Disease at the National Institute of Cancerology-Mexico: A 9-Year Experience

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Castleman disease is a diverse group of lymphoproliferative disorders with four variants sharing common morphological features on lymph node biopsy but having different etiologies, presentations, treatments, and outcomes. It is classified into unicentric and multicentric variants (MCD), the latter being a systemic disease with multiple lymphadenopathy, where the most common pathological finding is changes in plasma cell type. Based on etiopathogenic characteristics, it can be divided into Kaposi sarcoma herpesvirus (HHV8)-associated, idiopathic, and those related to an overlap with POEMS syndrome. This disease can occur in both HIV-positive individuals and immunocompromised individuals due to other causes. Here, we report our experience with a cohort of HIV-associated MCD cases from 2014 to 2022.

A total of 21 patients diagnosed with HIV and HHV8-associated MCD were observed at INCAN during the study period. The majority were men (95%), with a median age of 33y (range, 25 to 54y). The main histologic MCD type was plasma cells (52%), followed by the mixed variant (28%) and the hyaline vascular type (19%). Almost half of the patients were newly diagnosed with HIV (43%), and half of them had a CD4 cell count below 200 cells/mm3 at the time of MCD diagnosis (48%). Only 38% of known HIV patients were taking combination antiretroviral therapy (cART) at the moment of MCD diagnosis. The vast majority (95.2%) were HHV-8 positive by immunohistochemistry, and 66.7% were associated with Kaposi sarcoma.

About 52.4% of cases had another active opportunistic infection documented. The most frequently associated infection was syphilis (33.3%), EBV coinfection (23.8%), hepatitis B (14.3%), hepatitis C (4.8%), and CMV viremia (4.8%). 90% of patients developed B symptoms, 71% had fever, 100% had palpable adenopathy, 71% developed hepatosplenomegaly, and 19% had pleural effusion at presentation. In blood analysis, elevated LDH was present in 42.9%, hypoalbuminemia in 71.4%, thrombocytopenia (<100,000) in 35%, anemia (Hb < 8g/dl) in 20%, and renal failure (GFR <30 ml/min/1.73m2) in 10% of cases.

The treatment for Kaposi sarcoma at entry was a bleomycin and vincristine-based regimen in 52% of patients, with a median of 2 cycles (range 1-7). Five patients (23%) achieved a complete response without requiring additional treatment. Three patients (14%) with MCD attack criteria resolved systemic symptoms only with cART. Unfortunately, 14% of patients died before receiving treatment, and 38% were managed with "wait and watch" because they did not meet the criteria for MCD attack. The remaining 47% received treatment, with the most common regimens being R-Dox (20%), RCHOP (30%), CHOP (30%), Dox-liposomal (10%), and R-Etoposide (10%), with a median of 4 cycles (range 1-7). Eight patients (38%) received ganciclovir maintenance treatment until reaching negative HHV8 viral load. During the 9 years of follow-up, a total of 5 patients (23.8%) died, with 80% of them experiencing disease progression. The overall survival at 24 months was 77% (figure 1) , and the median survival was not reached at the time of evaluation.

In conclusion, our patients with HIV-associated MCD demonstrated similarities to global epidemiological data, with a predominant male population and onset between the fourth and fifth decade of life. The most common histology was the plasmacytic

cell subtype, and B symptoms were frequently observed. Hepatosplenomegaly was more prevalent than previously described, anemia and thrombocytopenia appeared to be more common hematological manifestations than reported. Due to a significant percentage of patients having uncontrolled HIV at diagnosis, and unknown KS at the same time of the diagnosis, further research is needed to establish a direct association with MCD and HIV pathophysiology in which clinical manifestations of the disease are diverse an lack specificity, and the prognosis varies greatly. Additionally, concomitant infections may play a role as activators of systemic inflammation and require active screening for MCD in patients with these characteristics. While Kaposi sarcoma treatment was effective in controlling both diseases in some cases, additional management may be necessary, with promising results seen with different treatments. Early intervention for cytokine release syndrome is essential to improve patient outcomes.

1.Blood 2020 Apr16;135(16):1353-64.
2.Nat Rev Dis 2021 Nov 25;7(1):84

Disclosures No relevant conflicts of interest to declare.

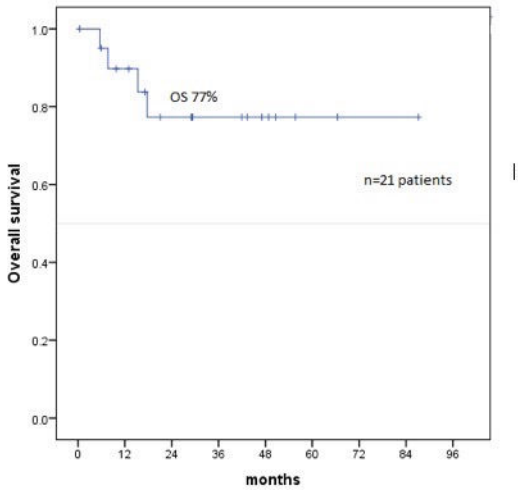

Figure 1

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