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Hematophagocytic Syndrome in a Patient Co-Infected With HIV: A Case Report

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ABSTRACT

Introduction: El síndrome hemofagocítico es una enfermedad grave caracterizada por un estado de hiperinflamación sistémica con sobreproducción de citoquinas. Puede responder a causas genéticas (primario) o desencadenarse por infecciones, fármacos, neoplasias o enfermedades autoinmunes. Con una mortalidad incrementada. Se presenta el caso de un paciente con VIH que desarrolló síndrome hemofagocítico en internamiento.

Case presentation: Presentamos el caso de un paciente masculino portador de Inmunodeficiencia humana, el cual desarrolló síndrome hemofagocítico con linfadenopatías y reporte de biopsia concluyendo la sospecha sindromática.

Conclusiones: Nuestro reporte de caso refleja la el abordaje clínico y bioquímico del síndrome hemofagocítico, en quien la realización de un diagnóstico histopatológico fue determinante para reflejar la causa de la enfermedad en cuestión, denotamos la importancia de la sospecha clínica en situaciones infrecuentes relacionadas con el paciente con inmunodeficiencia.

KEYWORDS: case report, hemophagocytic syndrome, immunodeficiency, HIV.

ARTICLE DETAILS

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CASE REPORT

Male, 29 years old, diagnosed with HIV in April 2023 with ARV treatment based on Triumeq that caused hepatotoxicity and was switched to temixclar (efavirenz 600-emtricitabine 200-tenofovir 245) in September 2023. The last viral load in October 2023 was undetectable and CD4 was 3207 ul.

Resided in Nuevo Leon and due to intense low back pain, MRI was performed which reported retroperitoneal lymphadenopathy, laparoscopic biopsy was performed with a diagnosis of tuberculosis reporting mesenteric lymph node with mycobacterial lymphadenitis, histological image compatible with infection by atypical mycobacteria, negative IHC stains for CMV and EBV, IHC remains do not show data of neoplasia CD68 (clone 4B5, membrane staining): positive, cytokeratin (clone AE1/AE3, membrane stain)negative, CD1a (clone EP 3622, membrane stain) negative, s100 (clone 4c4. 9, cytoplasm staining): negative. E started dotbal in intensive phase on September 3, 2023, concluding intensive phase on December 28, 2023. A control CT scan was performed with reduction of the size of the perivascular lymph nodes to 1 cm, normal thorax, hepatosplenomegaly 23 and 21.6 cm respectively. Negative tests for HBV, HCV and VDRL.

13.12.23 with hb 7.3, hypochromic microcytic, 5630 leukocytes, platelets 343 thousand, glucose 113, urea 18, creatinine 0.78, uric acid 6.9, cholesterol 130, Triglycerides 141, BT 0.33, AST 18, ALT 9. 8, GGT 68, FA 68, DHL 128, ALB 3.57, no DHE, iron 23, EGO normal, coprologic with mucus +, parasites -, SOH -, muscle fibers ++, fatty acids ++, starch ++, stool culture negative.

CXR 07.01.24: no pathological data, apparent radiopaque peribronchial circumscribed area in the lower bronchus. As well as apparent micronodules at the level of pulmonary window in multiple areas.

USG liver and spleen 06.02.24 splenomegaly rest without alterations.

CT simple and contrasted thorax and abdomen 09.01.24 cervical, retroperitoneal and mesenteric adenopathies, hepatosplenomegaly, probable lymphoproliferative disease.

Simple cranial CT scan 14.02.24 without evidence of alterations.

Reason for admission secondary to hyporexia, weight loss, myalgias and arthralgias, asthenia, fatigue, dysgeusia and liquid bowel movements without mucus or blood more than 10 times a day with orange color. Related in September when starting ARV treatment with temixclair, with severe anemia

and persistence of bowel movements, he was admitted to hospital on 06.01.24 with severe dehydration. On arrival at the emergency room with BP: 52/34, HR 99 FR 25, poor response to hydric resuscitation, starting vasopressor and showing metabolic acidosis requiring intravenous bicarbonate.

He was evaluated by the Infectious Diseases Department on 19.02.24 due to a high suspicion of hepatoxicity due to Dotbal, ruling out such compromise since there was no elevation of alkaline phosphatase or transaminases. However, at the physical examination she had cervical lymph node growths, with suspicion of pancytopenia and left cervical lymph node was biopsied on 02.20.24 to rule out lymphoproliferative process.

On 02.26.24 undetectable HIV viral load was collected again indicating virological control, but with CD4 count of 88 cells, so immunological failure was suspected, probably due to systemic disorder without ruling out opportunistic infection.

Bone marrow aspirate was performed on 02.29.24 due to persistent pancytopenia, finding in bone marrow smear (Figure 1) hemophagocytosis of cellular elements (erythroid precursors and myeloid series) integrating hemophagocytic syndrome due to splenomegaly, cytopenias, finding of hemophagocytosis in bone marrow and despite not meeting the 5 of 8 criteria, they support this diagnosis with hepatobiliary dysfunction and hypoalbuminemia. Treatment was started with etoposide on March 7, 24 and human immunoglobulin 5 grams per bottle on March 7, 24 administering 7 bottles in 1 hour every 24 hours for 2 days, as well as dexamethasone calculated according to sc 1.5 m2. The last cycle of etoposide was completed on 03.17.24.

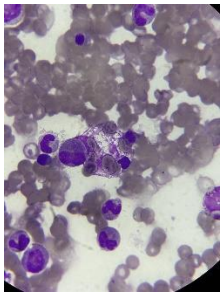

Figure 1. Hemophagocytosis of cellular elements (erythrocytes).

Peripheral blood smear report: 1% orthochromatic erythroblasts, marked anisocytosis, mild microcytosis.

DISCUSSION

Hemophagocytic syndrome (HS) is defined as a massive and ineffective stimulation of the immune system. This stimulation causes an increase in circulating inflammatory mediators and macrophagic tissue infiltration, phenomena responsible for the most dominant clinical manifestations 1 Although it is an unusual manifestation in HIV-infected patients, the phenomenon of hemophagocytosis has been found in up to 20% of autopsies of AIDS patients 16 This suggests that it may be an underdiagnosed manifestation linked to the disease, treatments administered or opportunistic infections, among others.

HS can be linked to genetic alterations that determine inadequate macrophage activation. When this occurs the disease usually manifests in childhood and is called primary HS.

On the other hand, HS can be triggered by infections, exposure to drugs, autoimmune diseases and neoplasms 1), and is called secondary when it occurs in this scenario. Viral infections are the most frequent trigger, and within these, those caused by herpes family viruses are responsible for more than 50% of the cases). Bacterial infections have also been identified as a trigger for HS, within these, tuberculosis has been blamed for up to 10% of the cases, mainly in patients with immunocompromise. In HIV-infected patients, HS can be triggered by opportunistic infections, with tuberculous infection and disseminated histoplasmosis standing out for their importance.

Our case describes a male patient with a history of HIV infection with undetectable viral load and last CD4 count of 88 cells, with a history of extrapulmonary tuberculosis also in antifungal treatment in the support phase; starting with hyporexia, weight loss, myalgia and arthralgia, asthenia, fatigue, dysgeusia and liquid evacuations without mucus or blood more than 10 times a day with orange color. With severe anemia and persistence of symptoms with data of hypovolemic shock, which merited the initiation of vasopressor, with elevated bilirubin and refractory pancytopenia even requiring the transfusion of 13 erythrocyte concentrates, fever, splenomegaly, as well as histological evidence of hemophagocytosis in bone marrow are concluded with 5 of 8 criteria according to HLH Criteria 2004, although it is known that HLH can be associated even with shock and cardiovascular collapse and acute kidney injury, liver dysfunction and coagulation disorders, this patient did not meet coagulation disorders since he was found without alterations in coagulation times.

The incidence of HLH is about 1 new case per 800,000 people per year and mortality remains high in HLH, reaching 40% in the entire population of patients with HLH, with support from the hematology service treatment was administered with etoposide + dexamethasone according to currently established treatment.

Another diagnostic classification, the Hscore, has been proposed for the diagnosis of reactive hemophagocytic syndrome (acquired forms of HLH). With items resulting in a final score of 90 to 250, higher scores correlate with a higher probability of HLH. The proposed optimal cutoff value was 169, which correctly classifies patients with HLH in 90% of cases.

● Includes 1 genetic criterion (presence of a genetic abnormality associated with inherited forms of HLH) and 8 clinical or biological criteria:
o fever
o splenomegaly
o Cytopenias (≥2 of 3 cell lines) with hemoglobin <9 g/dL, platelets <100 x 109/L and polymorphonuclear neutrophils <1.0 x 109/L
o Hypertriglyceridemia ≥3.0 mmol/L and/or hypofibrinogenemia ≤1.5 g/L
o Hyperferritinemia ≥500 mg/L
o Histologic evidence of hemophagocytosis on examination of bone marrow or lymph nodes.
o Low or absent NK (natural killer) cell activity
o Soluble CD25 ≥2400 U/mL

This explains the conduct taken with the case described, where an exhaustive search for opportunistic infections and pathologies associated with the deterioration, which were suggestive of hematologic damage, was performed, and the patient was maintained with immunomodulatory treatment and steroid therapy, with adequate response to it. We emphasize the importance of a good anamnesis, identifying primary factors and their related importance to emphasize the diagnostic-therapeutic approach.

-Appendix

Laboratory tests

	07.01.24	08.01.24	19.02.24	20.02.24	27.02.24	01.03.24
hb	6.2	7.4	9.2	8.3	5	
hcto	21	23.5	24.60%		12.1	
plaq	286 mil	262	74 mil	83 mil	42mil	
leucocitos	4800	5240	7.12	7.460	2.1	
Linfocitos			1.12	820		
Neutrófilos	3140		5.85	6490	1.25	
glucosa	116		70.5			
urea	19.8	14.6				
Creat	0.72	0.72	3.55			
BUN	9.25					
Na	129		125.8			
K	3.8		3.9			
Cl	97		97.2			
BT	0.31		11.50			2.02
BD	0.141		8.8			1.5
BI	0.17		2.67			
TGP	7.9		63.6			
TGO	18.2		24.6			
GGT			71			80.3
DHL	179	139				163
Retis		0.4%	3.02		2.7	
Antirubeola IgM		1.180				
Anti rubeola IgG		500				
albumina		2.6				
Fosfatasa alcalina			97.1			178
TP			16.1			

TPTa			49.6			
Fibrinogeno			461.			
Globulinas			5.6			
ferritina					811	

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