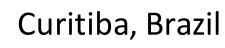
II ASBAI-BRAGID MEETING 2016

Case Presentation

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History

- EDSA, 12 years old boy.
- First child with no consanguineous parents.
- No family history of immunodeficiency.
- Gestacional history and childbirth: normal.
- Vaccination up-to-date with no adverse reactions.

History

8 years-old he had met 5 criteria for HLH: Fever

Splenomegaly Cytopenia (anemia and neutropenia) High ferritin (2,434 ng /mL) High triglycerides (312 mg/dL).

Positive IgM for Epstein-Barr virus (2012) Quantitative PCR - 142 viral EBV copies/ul (2012)

He was treated with Dexamethasone, antibiotics and Acyclovir.

He remained asymptomatic for 3 years.

History

• 12 yrs (2015)

Starts with 4 months of intermitent fever, fadigue, sore throat, diffuse abdominal pain and persistent neutropenia.

He was positive serology for Epstein-Barr virus (IgM).

Since then, four admission by fever without source.

Physical exam

Weight=89Kg (90th P) Height: 149 (50th P) BMI=40 T=36.2°C

HR: 107 bpm RR: 22 ipm BP: 110x63 mmHg

Compromised clinical condition.

Cutaneous and mucous pallor.

Abdominal reddish stretch marks and central obesity.

No lymphadenopathies or hepatomegaly.

Laboratory investigation

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Hb=9.2g/dL, MCV=77fl, Pla=178,000 cells/uL.
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Leukocytes = 1890 cells/uL
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Neutrophils = 1% (17 cells/uL), Lymphocytes = 54%, Monocytes = 45%.
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Normal liver function tests

Positive Rubella IgG / IgM negative (March/16)

EBV VCA-IgM=43.2 U/ml EBV Anti-EBNA IGG=80.1 U/ml (January/16) Negative IgM for EBV (March/16)

Abdominal ultrasonography showed liver steatosis and homogeneous splenomegaly (16cm).

Laboratory investigation

Antibodies (mg/dL)		January 2016			March 2016			
IgA		235,8			319,4 (H)			
IgM		1485 (H)			683,2 (H)			
IgG			1469 (H)			2327 (H)		
IgE (IU/mL)			<25			< 25		
	Lymphocytes	CD3	CD4	CD8		CD19	CD16/56	
January	1760	1408	292	809		42	25	
March	2243	1883	560	1081		0	340	

Absolute lymphocytes and subset counts (cells/uL)

Laboratory investigation

HIV negative

Bone marrow immunophenotyping : 0.34% precursor cells of the myeloid lineage. Intensely altered pattern of neutrophil maturation. Maturation arrest in promyelocytes / myelocytes.

Biopsy Neutrophils and red blood cells inside macrophages.

