

# 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 intermittent fever, fatigue, 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 (90<sup>th</sup> P)      Height: 149 (50<sup>th</sup> 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

Hb=9.2g/dL, MCV=77fl, Pla=178,000 cells/uL.

Leukocytes = 1890 cells/uL

Neutrophils = 1% (17 cells/uL), Lymphocytes = 54%, Monocytes = 45%.

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.



