

Abstract –

Presumed Disseminated Mycobacteriosis and severe lymphopenia

K.F.R, admission age: 1y 3 mo, Brazillian, White

Patient referred to our institution with a history of fever and hepatosplenomegaly (10 cm and 12 cm) without improvement using ceftriaxone. On admission, she presented fever, hepatosplenomegaly, severe anemia and a respiratory disease. She was internalized and started an investigation to make a diagnosis. Her first exams showed pancytopenia, hypergammaglobulinemia and elevated LHD and C - reactive protein. While we were discarding leukemias, HIV, histiocytosis and Kalazar, she was getting worse.

Previous: Many otitis and two pneumonias in the first year

Parents: Not relatives, no past history of immunodeficiency

Exams:

Negative ELISA: CMV, Toxoplasmosis, Parvovirus, HIV, Epstein-barr, Leishmania, and Brucellosis.

Negative PCR: Leishmania

Protein electrophoresis and immunofixation electrophoresis: normal

Cultures: negative (blood, hepatic, bone marrow)

Myelogram: Moderate erythroid hyperplasia

Liver Biopsy: Focal cholestasis. Kupffer cells hypertrophy and hyperplasia, rare microgranulomas with giant cells, without apparent necrosis.

Bone marrow biopsy: Granulomatous inflammation (tuberculoid) anti CD 68 positive and anti S100 negative.

NBT: normal

PPD: negative

Following these results, mainly biopsies, we decided to treat as disseminated atypical mycobacteriosis, even though no bacillus had been isolate.

Once the treatment started, she improved her outcome and fever disappeared in fifteen days. However, her lymphocytes number maintained low levels, despite increasing in platelets and red cells, and for such reason she also received SXT. The

patient was dismissed after 3 months internalized, with SXT and anti-mycobacteriosis medications.

She has been keeping the treatment, reducing slowly hepatosplenomegaly (5cm and 3 cm) and improving her general status. Her lymphocyte number improved in relation to that prior to treatment, but she has maintained levels below the normal for her age. Her total and specific immunoglobulins are normal (anti-pneumo not available).

	CD4	CD8	CD4/CD8
06-2004	232 (79.5%)	40 (13.6%)	5.80
08-2004	630 (38.5%)	953 (58.2%)	0.66
01-2005	571 (64.9%)	229 (26.5%)	2.49

	IgG	IgM	IgA	IgE
04-2004	1068	173		
08-2004	926	138	62	
10-2004	1047	211	73	288

	LDH	CRP
5/11/2004	799	46.9
5/27/2004	3607	241 → Riphampicin reaction
7/14/2004	392	2.4

Points:

- 1- Is it really Mycobacteriosis? (*M.bovis* or others)
- 2- Lymphopenia: primary or secondary?
- 3- Defects in IL12-INF pathways?
- 4- How long should she receive treatment?
- 5- What further investigation could we perform to elucidate this patient better?

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