

# *Case Presentation*

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# *Case Presentation*

- 37 years-old-female
- Crohn's disease
  - Terminal ileitis (colonoscopy, CT enterography)
- Treatment:
  - Budesonide 9mg + 6 MP 75 mg

# *Hospitalization*

- Fever up to 38.3° for 7 days
- Without other symptoms
- Physical examination – unremarkable
- Medications:
  - Budescan 9mg/d
  - 6 MP 100mg/d

# *Presentation Labs*

- WBC- 1570 k/micl
  - Neut- 800 k/micl
  - Hg – 11.9 g/dl (MCV 98)
  - Plt- 93 k/micl
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- Coagulation- normal
  - CRP 2.248
  - Alb 4.3 g/dl
  - Globulin 2.8 g/dl

- Bil total 1.89 mg/dl  
(Direct 0.52)
  - ALP 114 U/L
  - GGT 140 U/L
  - ALT 127 U/L
  - AST 114 U/L
- 
- LDH 1032 U/L
  - CPK 62 U/L
  - Iron 46 mcg/dl
  - Transferrin 174 mg/dl

# *Work-Up*

- Chest X ray - normal
- Abdominal US:
  - Mild fatty liver
  - Mild splenomegaly

# *Problem List*

- Crohn's disease – Tx with budesone and 6MP
- Fever
- Pancytopenia
- Elevated liver enzymes

# *Work- Up*

- Broad infectious serology – negative
- CMV IgM positive
- CMV PCR 16500 cop/ml
- Treatment for CMV infection
  - Gancyclovir

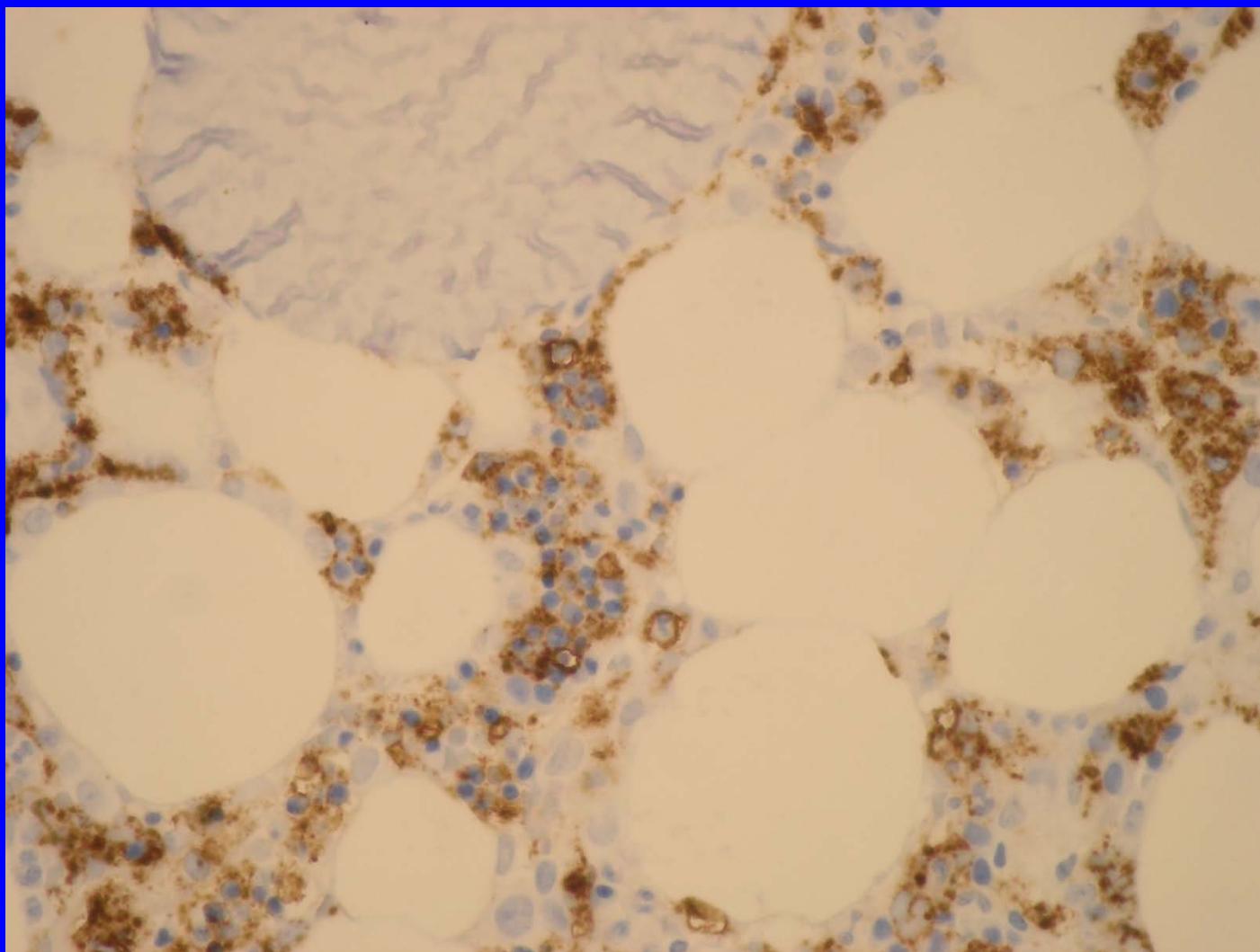
# *Chest CT*



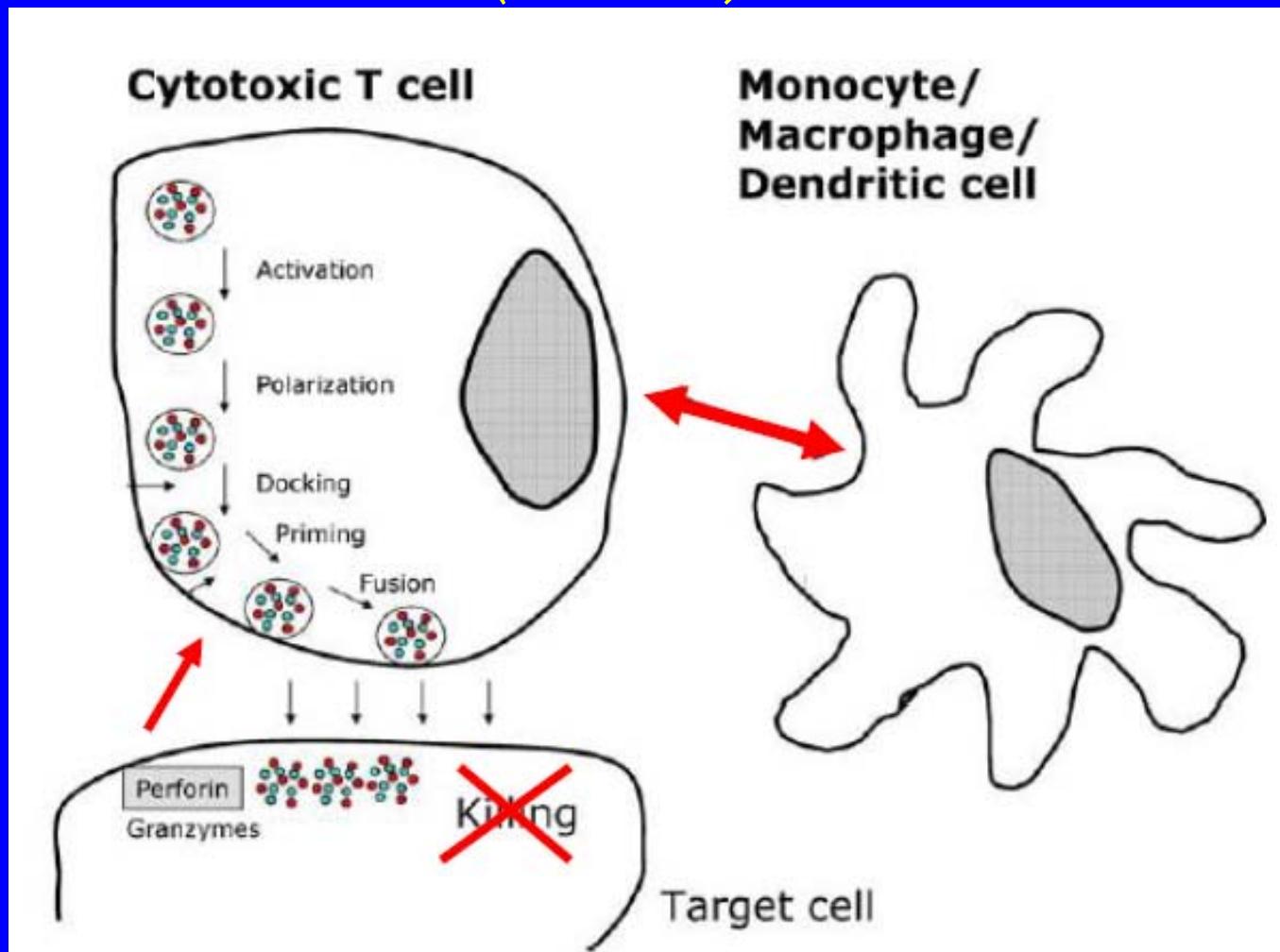
# *Follow - Up*

- Fever continues
- Worsening of pancytopenia
  - WBC – 700 k/micl ,Neut- 300 k/micl
  - Hg 7.6 g/dl
  - Plt 34 k/micl
- No signs for hemolysis
- Treatment as neutropenic fever:
  - Broad spectrum ABX
  - GCSF

# *Bone Marrow*



# *Hemophagocytic Lymphohistiocytosis (HLH)*



# *Hemophagocytic Lymphohistiocytosis (HLH)*

- Primary HLH - genetic
- Secondary HLH
- Clinical findings-
  - Fever, splenomegaly, cytopenia, hepatitis and neurologic symptoms
- Prognosis- 50-70% 5y survival with TX

Kimberly Rismaa and Michael B. Jordan, Curr Opin Pediatr 2012

# *HLH- Diagnosis*

- Fever
- Splenomegaly
- Cytopenia (at least 2 cell lines)
- Fasting TG > 265 mg/dL, and/or Fibrinogen < 150 mg/l
- Hemophagocytosis in BM, CSF, or LN
- Decreased/absent NK cell activity
- Ferritin > 500 ug/l
- Soluble CD25 (IL2) > 2400 U/ml

# *HLH- Treatment*

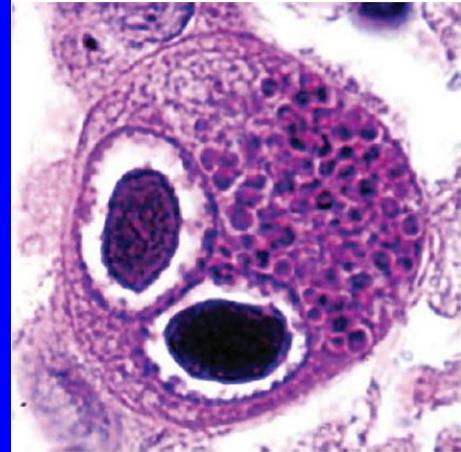
- Discontinuing of immunosupresion
- Direct treatment for the cause
- Chemotherapy
- Allogeneic stem cell transplantation

Jordan MB, Allen CE, Weitzman S, et al. . Blood 2011

# *HLH and IBD*

- Case reports of HLH and IBD (Crohn's>>UC)
- Most frequent with immunomodulators Tx
- Induced by infection
  - EBV
  - CMV
  - Histoplasmosis
  - TB

# *CMV and IBD*



- Colitis can mimic IBD
- Associated with severe or steroid refractory disease
- UC>>Crohn's

# *Back To Our Patient*

- Abdominal US:
  - Hepatosplenomegaly
- Ferritin- 7000 µg/l
- TG-325 mg/dl
- NK activity- 46%
- Soluble IL2- 4013 U/ml (300-2000)

# *Follow-Up*

- Treatment:
  - Gancyclovir → Foscarnet
  - IVIG
  - Dexacort
  - VP16

# *Follow - Up*

- Clinical improvement
- Blood count- improved
- CMV PCR-negative
- Bone marrow:
  - Minimal hemophagocytosis
  - Hematopoietic improvement

**Thank you**