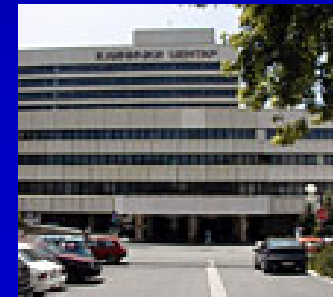




Clinical presentation

ESIM, 6-13 Sept 2009

Clinical Center of Serbia
Dragana Mijač, MD



DJ 47 yrs, female

HPI: Abdominal cramps, diarrhea,
face and lower leg edema, weight loss (6 kg /1mo), fatigue

PMH: Schistosomiasis 6 years ago (Africa)
4 miscarriages

Ph. exam.: BMI 17,6 kg/m², eyelids edematous,
conjunctiva anemic, pretibial pitting edema

Routine lab

- Hgb 108g/l, MCV 75.1fl, Fe 6.8 μ mol/l, TIBC 40.6 μ mol/l, ferritin 4.7 μ g/l
- AST/ALT 60/55U/l
- Chol. 2.91 mmol/l
- CK139 U/l, LDH 406 U/l
- Alb. 18g/l TP 40g/l

Differential Diagnosis

Infection: Schistosomiasis, histoplasmosis, amebiasis, *C. difficile*, HIV, TB, HBV, HCV etc.

Other: Chronic pancreatitis, hyperthyroidis, Crohn's disease, lymphoma, sarcoidosis, PBC, PSC, celiac sprue, polymyositis, intestinal lymphangiectasia etc.

Lab exam and procedures

HIV, HCV, HBV, Mantoux, TSH, T3, T4: normal

Immunologic tests:

antigliadine-antibody, antiendomysial-antibody, AMA, ANA,
antiJo1, ASMA, neg **Anti-parietal Ab + (1:320)**

Stool tests

ova, parasites: negative

culture (includ. *C. difficile*): negative

occult blood, steathorea/fat, F-calprotectin: negative

Fecal excretion alpha 1 antitripsine: 483.6 ml/24h (<24)

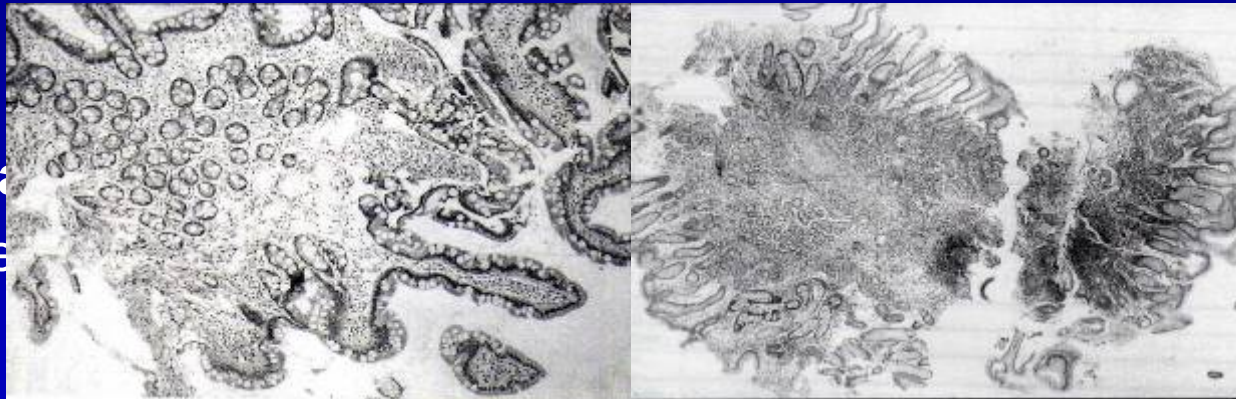
significant exudative enteropathy

Lab exam and procedures (cont'd)

Small bowel enema: flocculation

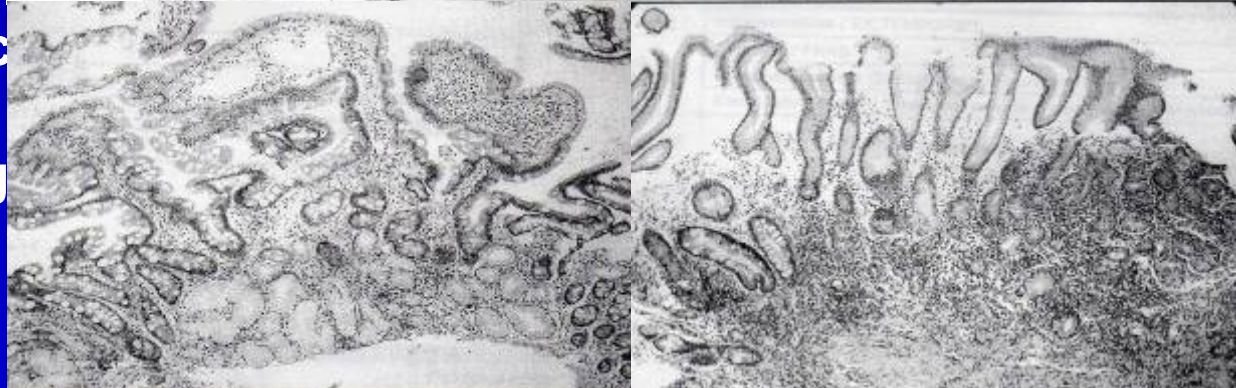
Upper endoscopy with enteroscopy and polymorph biopsies:

Double barium
nonspe



Colonosc

Lymphog



normal

Lab exam and procedures (cont'd)

Muscle biopsy: endomysial lymphocytic infiltration

Electromyography: muscle lesions consistent with sec. myopathy

Needle liver biopsy: very mild nonspecific changes

Diagnosis

Exudative enteropathy of unknown etiology
with secondary miopathy and atrophic gastritis

Tx: Gluten free diet
 Lactose free diet

Two years later

Symptoms: diarrhea (8 to 10 stools daily), vomiting, fatigue

Ph. exam.: malnourished (BMI 16.6), T 38C, pale, with severe leg edema, disseminate HSV infection, left lobe focal pneumonia

Abnormal lab values:

Hgb 92g/l, MCV 74.1fl, Fe 2.8 μ mol/l, TIBC 52.3 μ mol/l, ferritin 6.3 μ g/l,

AST/ALT 66/58 U/l

Alb. 13 g/l TP 34 g/l

Chol. 2.08 mmol/l

LDH 749 U/l, CK 447 U/l

- HSV1 IgM + (1:360)

Chest x-ray: focal pneumonia

Tx: Ceftriaxone 2g i.v. for 10 days and Aciklovir 800 mg i.v. tid for 10d

Two years later (cont'd)

- Previous diagnostic panel was performed, with similar findings in: fecal, microbiological, immunological, endoscopic, microscopical findings

- **Test for common variable immunodeficiency (CVID):**

IgG low

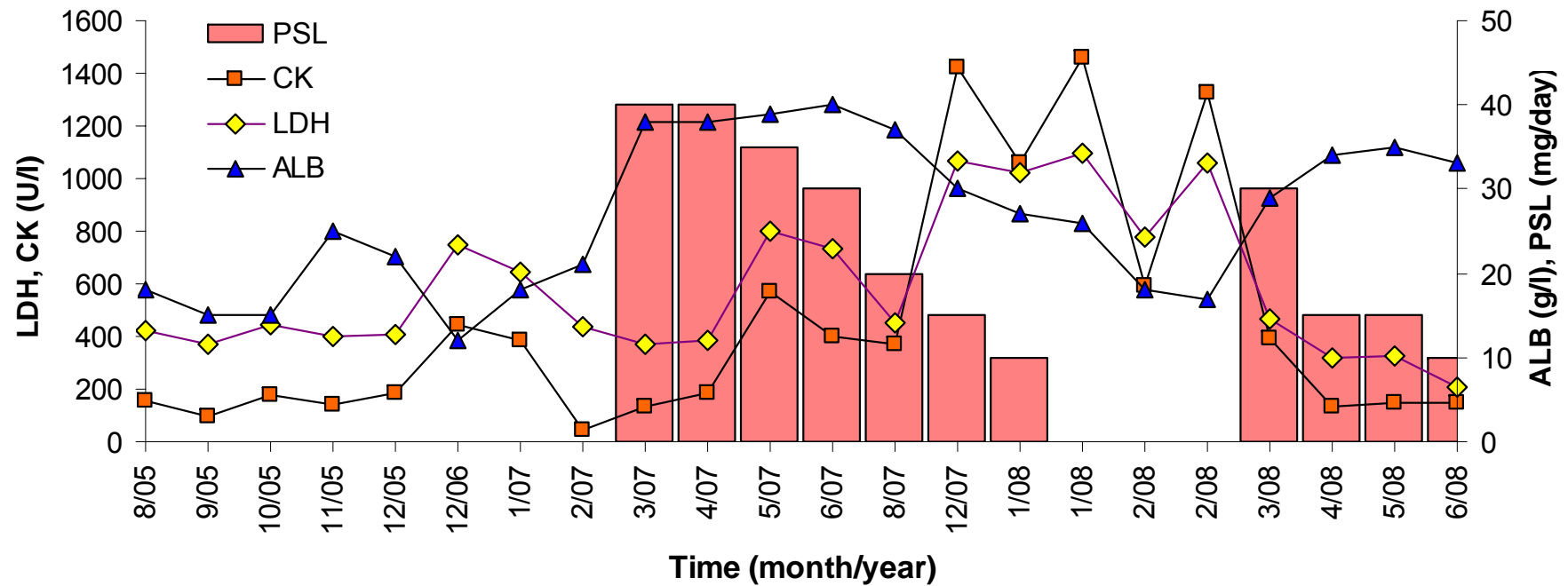
Lymphocytic proliferative answer to mitogen was lower (4568 cpm, compared to two controls - 29090 and 26178)

Definitive diagnosis

Autoimmune protein-losing enteropathy with atrophic gastritis and sec.myopathy

Tx: Prednisolone 40 mg/day + Ca + vit D

Follow up



PSL – Prednisolone
CK – Creatine kinase
LDH – Lactate dehydrogenase
ALB – Albumin

Autoimmune protein-losing enteropathy

- Rare cause of intractable diarrhea in infancy
- A variant with onset in adulthood has recently been described
- Condition with adult onset chronic diarrhea, malabsorption, complete lack to response to gluten free diet or other dietary exclusions with small intestinal histologic features.^{1, 2}
- The immune mechanisms underlying adult AIE are unclear and the experience in the management of adult AIE is very limited ³

¹ Akram et al. , *Clin Gastroenterol Hepatol.*, 2007

² Aoki et al., *J Gastroenterol.*, 2002

³ Freeman, *World J Gastroenterol.*, 2008

Autoimmune protein-losing enteropathy

Therapy (long-term immunosuppression)

- Corticosteroids
- Azathioprine
- Cyclosporine A
- Infliximab
- Tacrolimus
- Cyclophosphamide

Thank You

