Is it really Crohn's?

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Introduction

- Crohn's disease – systemic inflammatory disease
- Complications involving multiple organs
- Newly described complications (Cogan syndrome, Sweet syndrome, etc.)
- A series of 3 clinical cases depicting rare complications and "acceptable" errors in diagnosis of Crohn's disease

1st Patient – Mrs. G 19y old

- August 2012: abdominal pain investigated by upper GI endoscopy showing granulomatous gastritis negative for H.pylori infection. No follow-up
- Actually: abdominal pain and fever after ingestion of NSAIDs
- No associated diseases
- No familial history for autoimmune diseases, cancer, cardiovascular diseases, diabetes
- Non-smoker

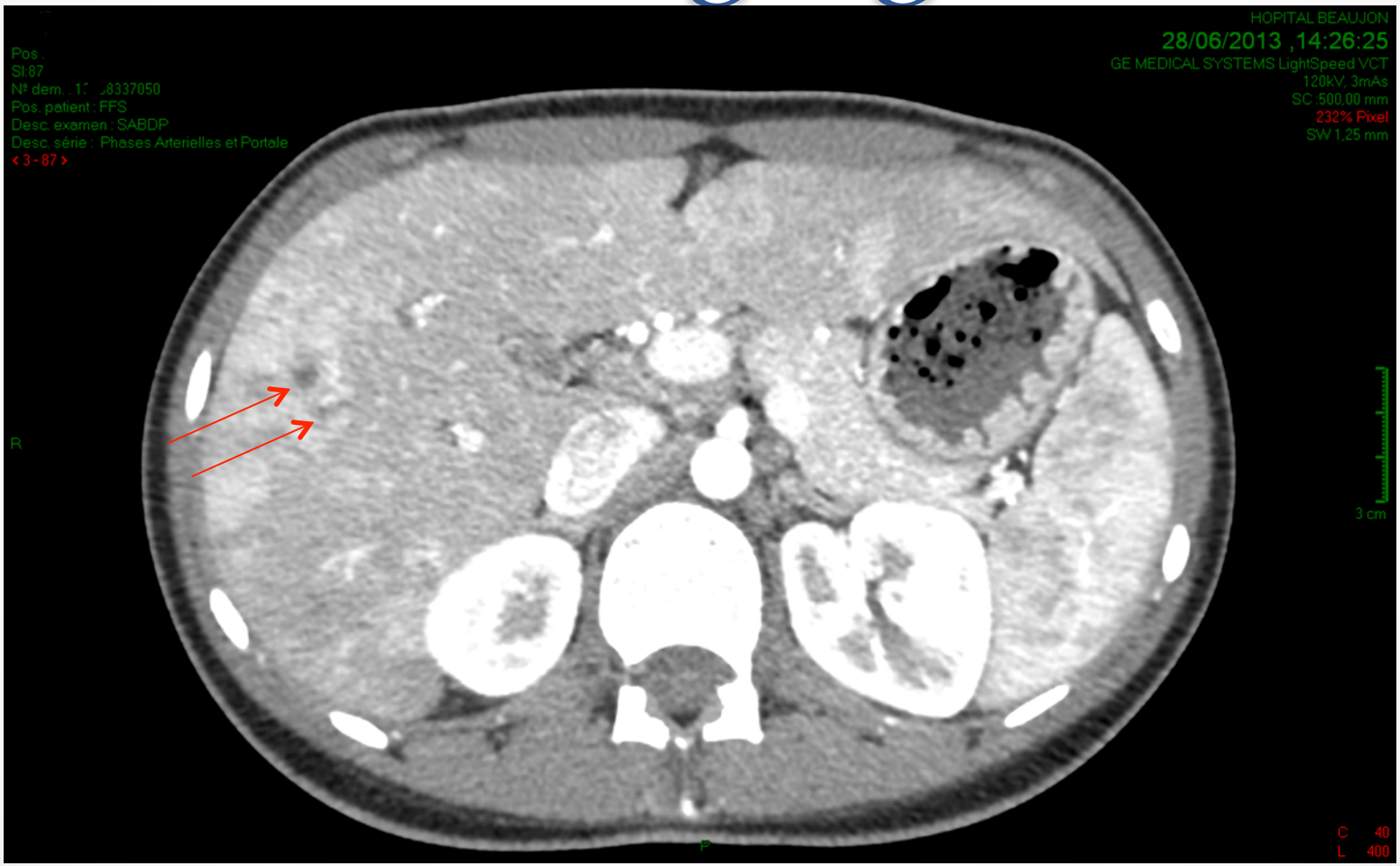
Clinical examination

- TA 125/90 mmHg; FC 98/min; 39°C
- Palpatory right upper quadrant pain
- Hepatomegaly
- No jaundice
- Negative Murphy probe
- Normal pumonary examination
-

Laboratory tests

- CBC: leucocytosis
- Inflammatory tests: CRP 90 mg/l
- Normal transaminases, mild cholestasis
- Negative blood cultures
- Negative urinary cultures
- Normal ACE, pulmonary CT scan, B2-mycroglobulin, LDH, ANA
- Negative Quantiferon TB

Imaging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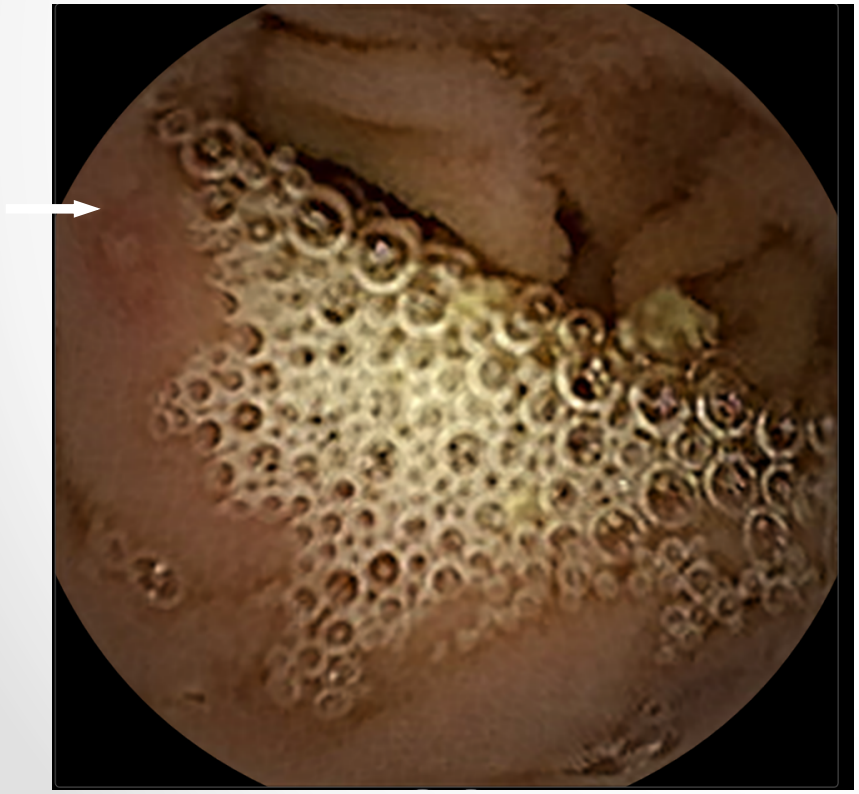
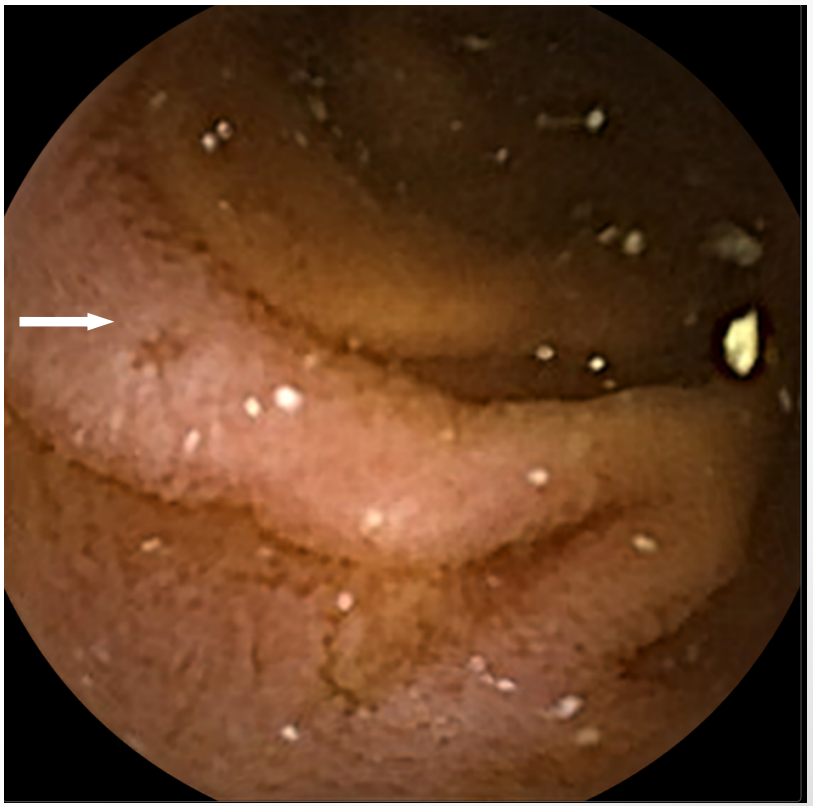
Evolution

- FNA of hepatic abscess
  - PMN ++
  - Negatve bacteriology
  - Negative parasitology
  - Negative fungus tests
- No amelioration after 2 trials of empiric antibiotic therapy

Endoscopy

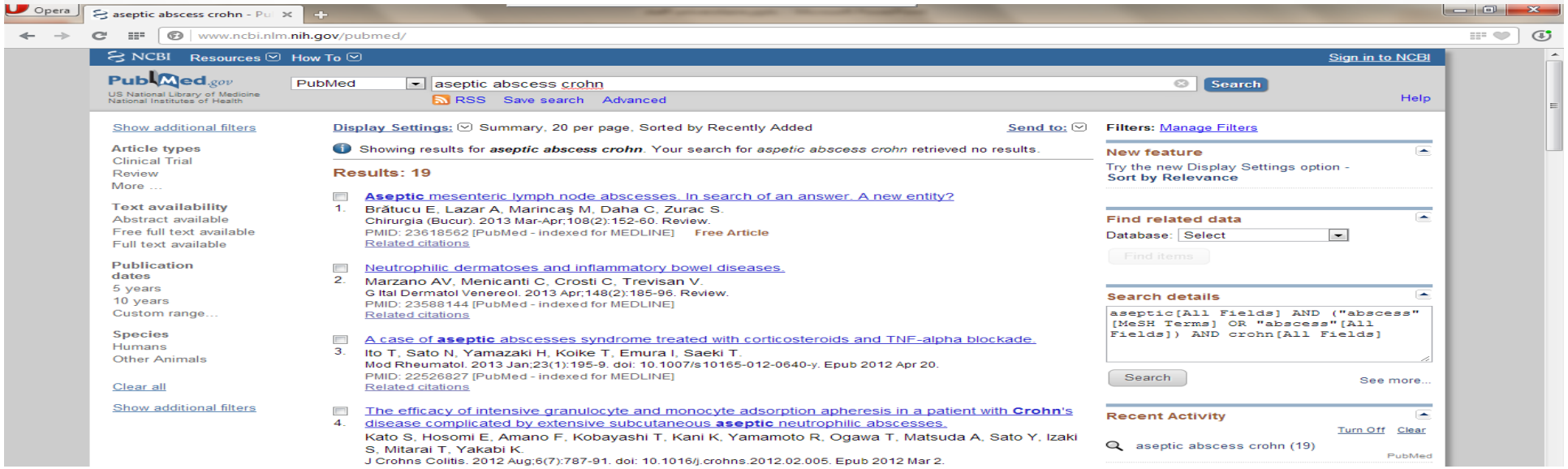
- Upper GI endoscopy – granulomatous gastritis
- VCE – multiples aphtoid lesions from duodenum to ileum





What is this?

- Complication of Crohn's disease?
- Another associated disease?
- What treatment?

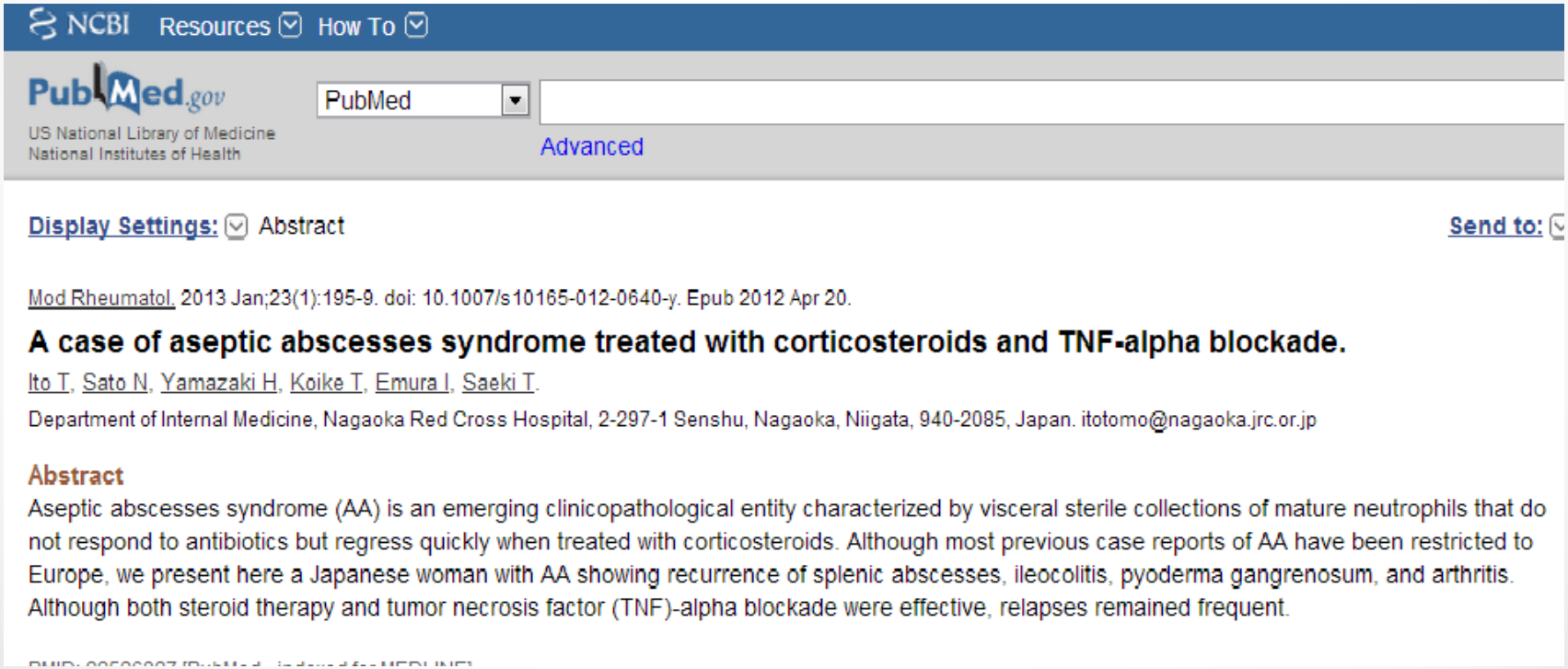


Diagnosis

- ***Aseptic abscesses associated to extended ileal Crohn's disease***

Treatment?

- The same as Crohn's disease – corticoids, immunomodulators, antiTNF



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Mod Rheumatol. 2013 Jan;23(1):195-9. doi: 10.1007/s10165-012-0640-y. Epub 2012 Apr 20.

A case of aseptic abscesses syndrome treated with corticosteroids and TNF-alpha blockade.

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Abstract

Aseptic abscesses syndrome (AA) is an emerging clinicopathological entity characterized by visceral sterile collections of mature neutrophils that do not respond to antibiotics but regress quickly when treated with corticosteroids. Although most previous case reports of AA have been restricted to Europe, we present here a Japanese woman with AA showing recurrence of splenic abscesses, ileocolitis, pyoderma gangrenosum, and arthritis. Although both steroid therapy and tumor necrosis factor (TNF)-alpha blockade were effective, relapses remained frequent.

Treatment

- Treated with Budesonide 9mg/day and Azathioprine for mantainance
- Remission of symptoms after 2 days
- Normal imaging at 3 months

2nd Patient – Mrs.F 27y old

- Diagnosis of colonic Crohn's disease in 2007
- Non-smoker
- No associated diseases
- No familial history for autoimmune diseases, cancer, cardiovascular diseases, diabetes
- 2007 – 2010 – *5ASA + corticoids for several mild-to moderate flares*
- Sept 2010 – severe flare→ CS; introduction of AZA 150mg/day; eritema nodosum and diarrhea after decreasing corticoids
- Dec 2010 combotherapy IFX 5mg/kgc + AZA

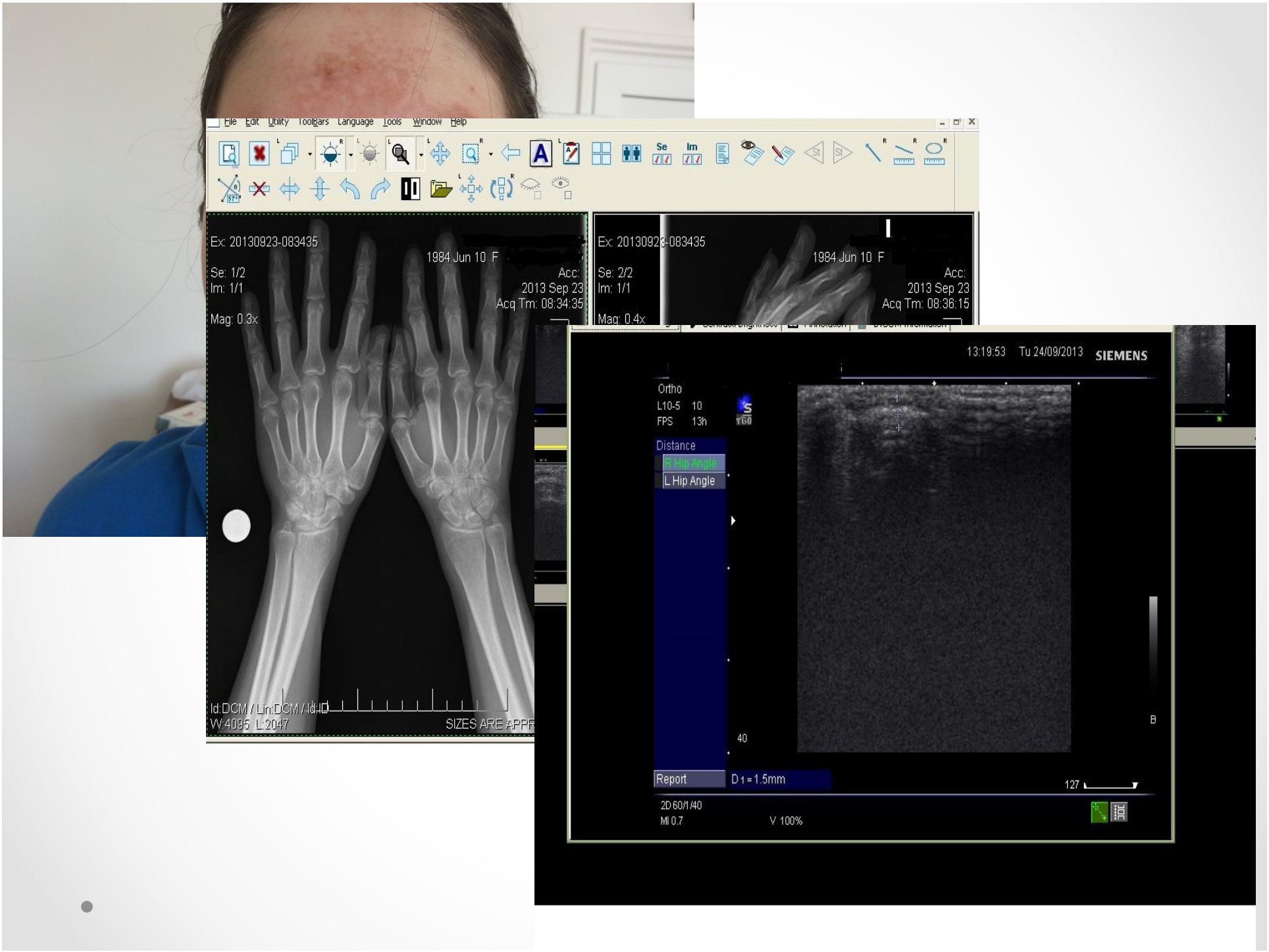
Evolution

- 2011 – dec 2012 remission under combotherapy
- Dec 2012 progressive periferic arthralgias two weeks after IFX perfusion

  Laboratory: CRP 4.95ng/dl, ESR 52mm/h
- Feb 2013 bicitopenia with mild macrocytic anemia and leucopenia (reduction of AZA to 100mg/zi + folic acid 5mg/zi)

Evolution

- July 2013 – persistance of symptoms (peripheric arthralgias responsive to Dexametasone treatment)
  - ***Type II arthropathy associated to IBD?***
- New symptom – facial erythema
- Bicytopenia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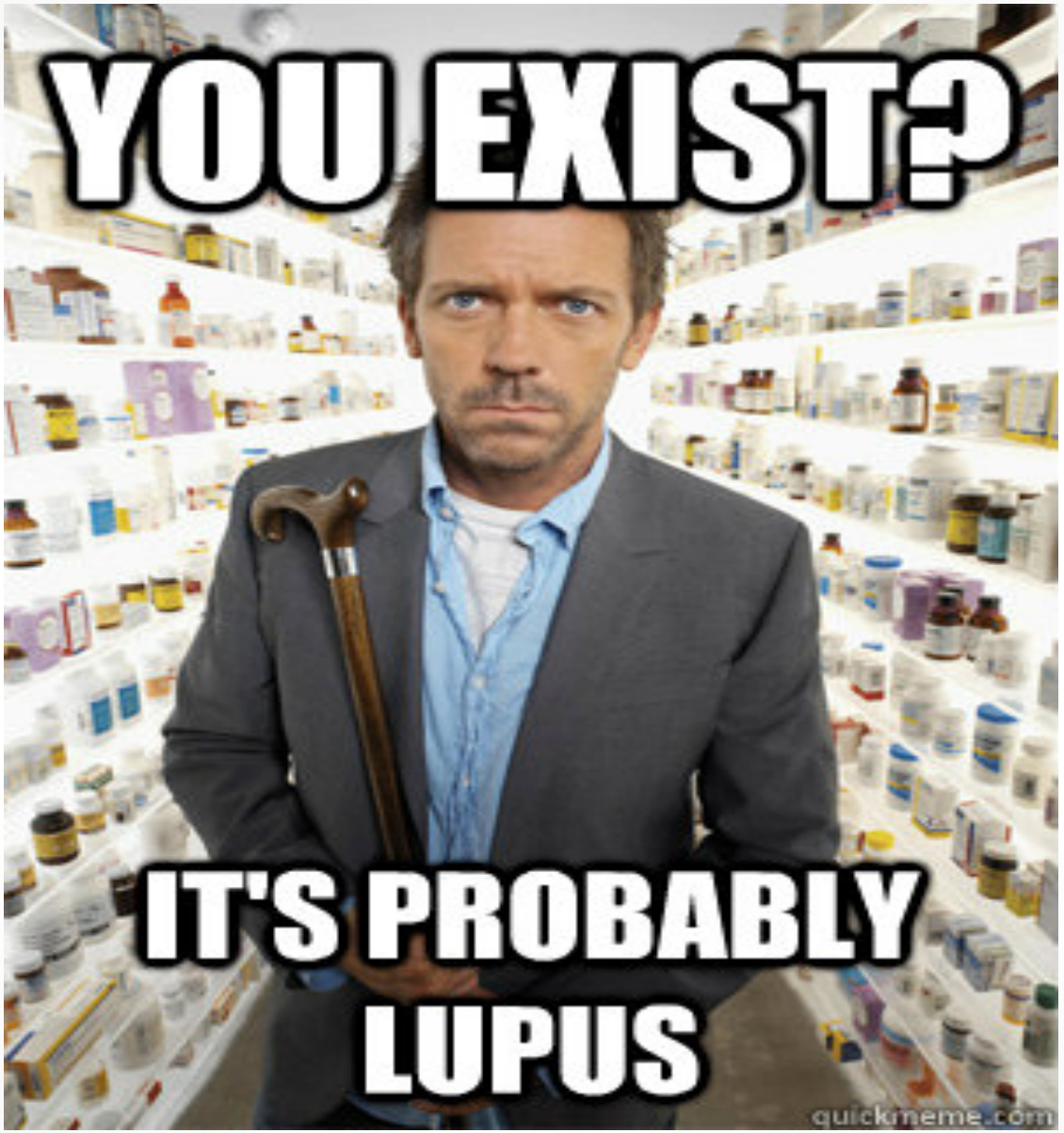
MI 0.7
V 100%

Laboratory

- CBC: ***leucopenia 3350/mmc*** , ***Hb 10.2g/dl***, PLT 271000/mmc, Fbg 343mg/dl, ***ESR 46mm/h***, ***CRP 5.05mg/l***
- *ADNds antibodies 21.5 UI/ml* (borderline)
- ***ANA 1/1280*** (N≤1/80) (Sp 79-100%)
- CCP antibodies 2.6 RU/ml (N<5)
- ***Anti-hystone antibodies 120 U/ml*** (N<40)


YOU EXIST?
IT'S PROBABLY LUPUS
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Diagnosis

- ***TAILS – TNF alpha induced lupus-like syndrome***
- Treatment???
  - No recommendations available

- Switch to Adalimumab
- Good evolution of arthropathy and mantainance of remission

…at least for now

3rd Patient – Mr. G 66y old

- Medical history :

Vaquez disease since 1996

Myocardial infarction with active stent June 2013

- Surgical history:

Appendectomy at 30y old

- Smoker – 5 PA

Evolution

- Jan 2012 : diarrhea, abdominal pain, fever, oral aphthosis, erythema nodosum – spontaneous remission

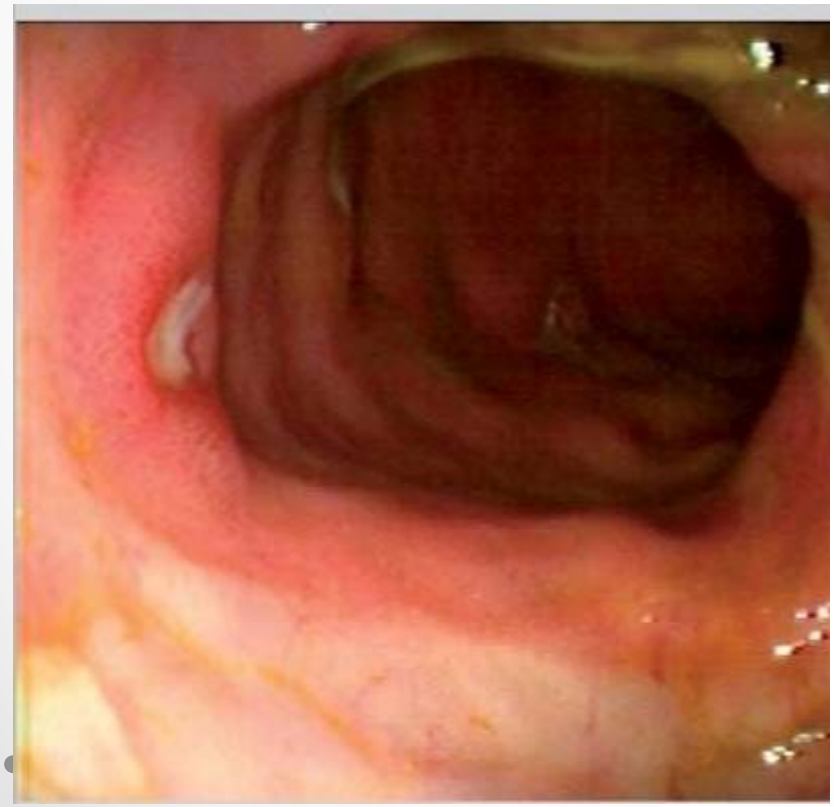
- Nov 2012 - asymptomatic
  - Ileocoloscopy : ileal ulcerative lesions
  - Upper GI endoscopy: normal
  - Treatement with 5-ASA : inefficient

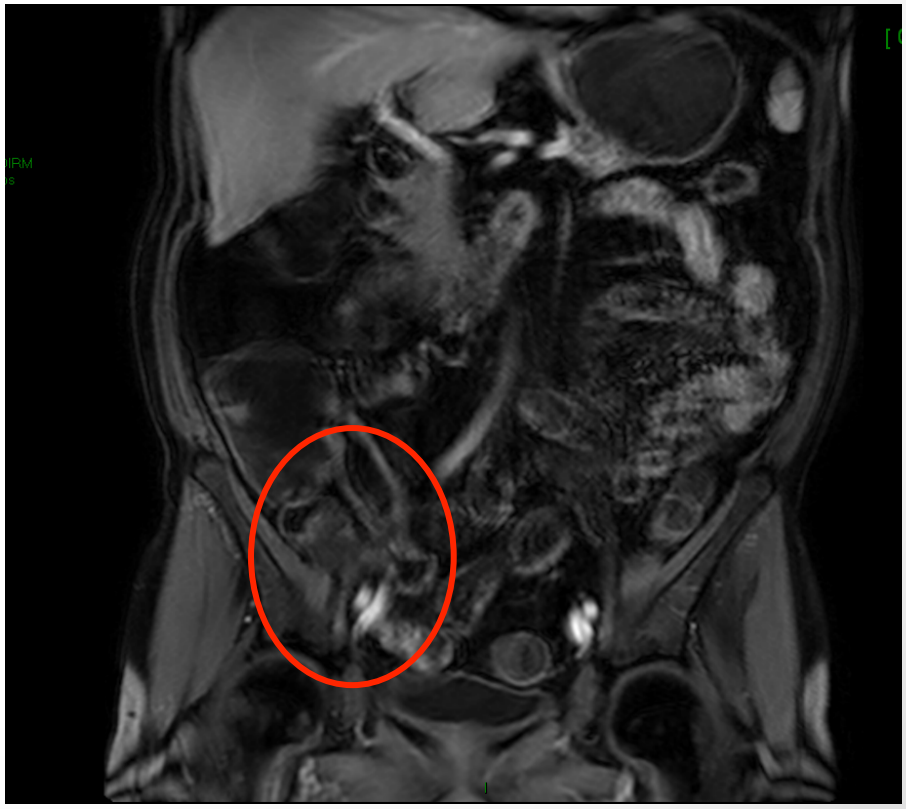
    Budesonide 9mg/j : inefficient

- Aug 2013 : relapse of abdominal pain, diarrhea, oral aphtoid lesions , episodes of vesperal fever

Investigations

- Ileo-colonoscopy + enteroIRM with macroscopic aspect of ileal Crohn's disease
- Histologic exam – suggestive for Crohn's disease(oct 2013)



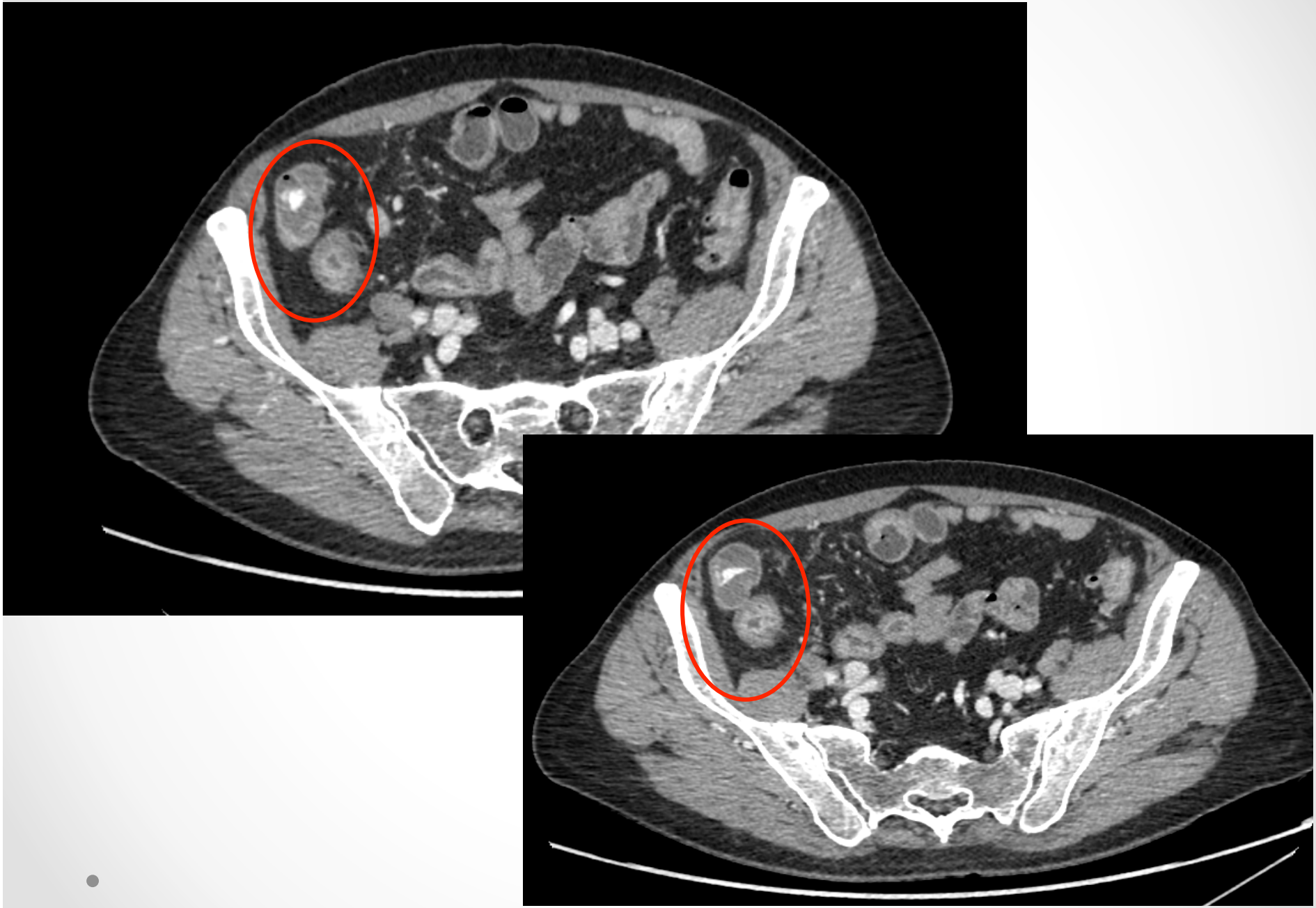


Evolution

- Oct 2013 : decision to start Adalimumab 160-80-40 immediately efficient
- Jan 2014 : loss of 3Kg, fever, abdominal pain, 2 to 3 stools/day with blood, 2 oral apthoid lesions

Evolution

- Brutal decrease in Hb level from 13 to 10g/dl
- CRP=200mg/dl
- Fever= 39,5 Celsius
- Tachycardia = 110bpm
- Negative blood cultures



What to do?

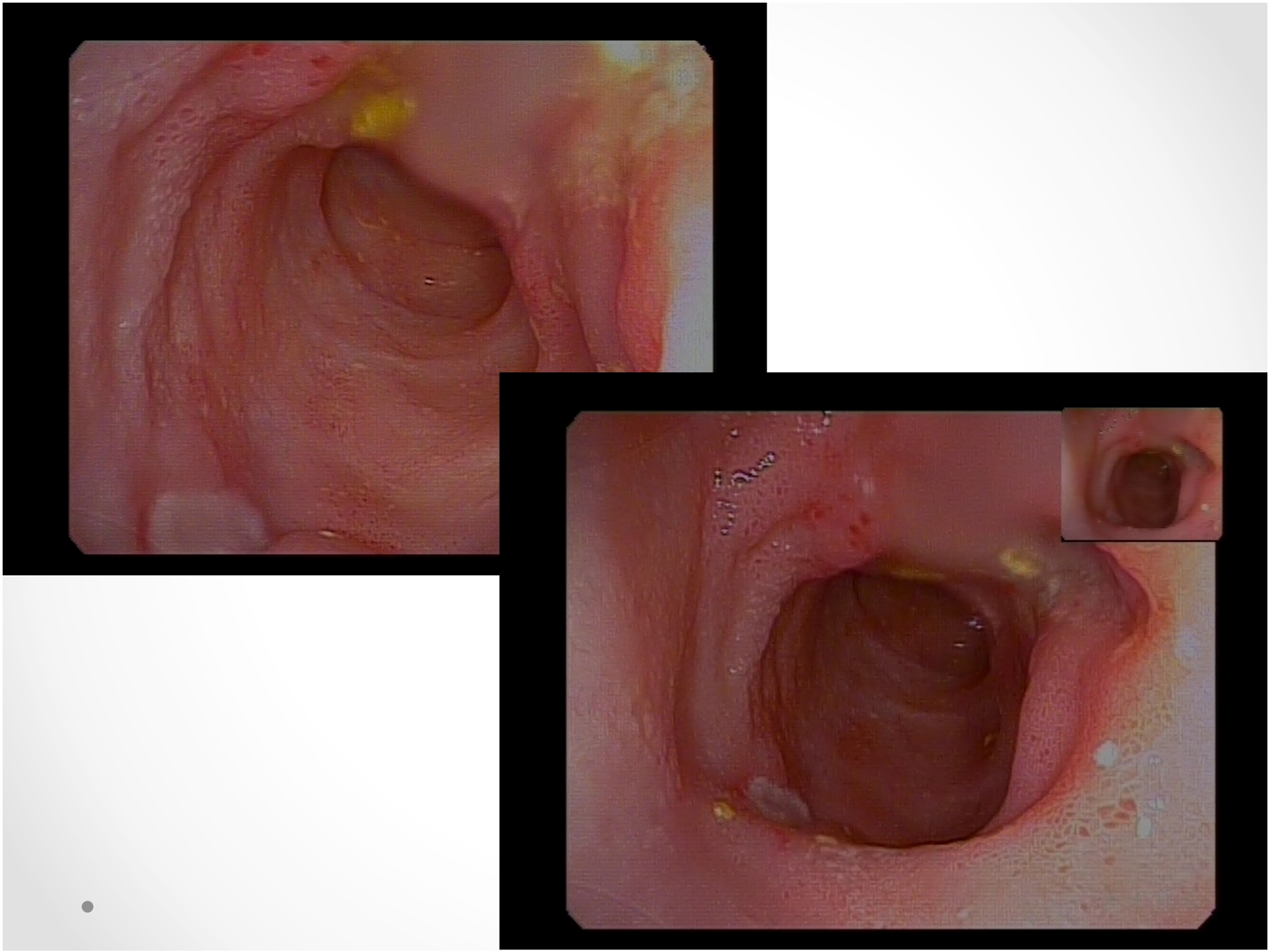
- Transfusion of red blood cells
- Treatement with bolus Methylprednisolone
    - Normalisation of stools (no blood), stablisation of CBC
- Relapse of fever and digestive symptoms when switching to oral systemic corticoids

What to do?

- Mars 2014 – coelioscopic ileo colonic resection with ileo-colostomy

- Peroperative period – relapse of fever and digestive bleeding by ileostomy
- Infectious causes exhaustively excluded
- Endoscopy by stoma showed profound ulcers over 15-20cm from stomial oriffice
- Restarted on i.v. Methylprednisolone with remission of bleeding

  - ***VERY early Crohn relapse in ileostomy???***

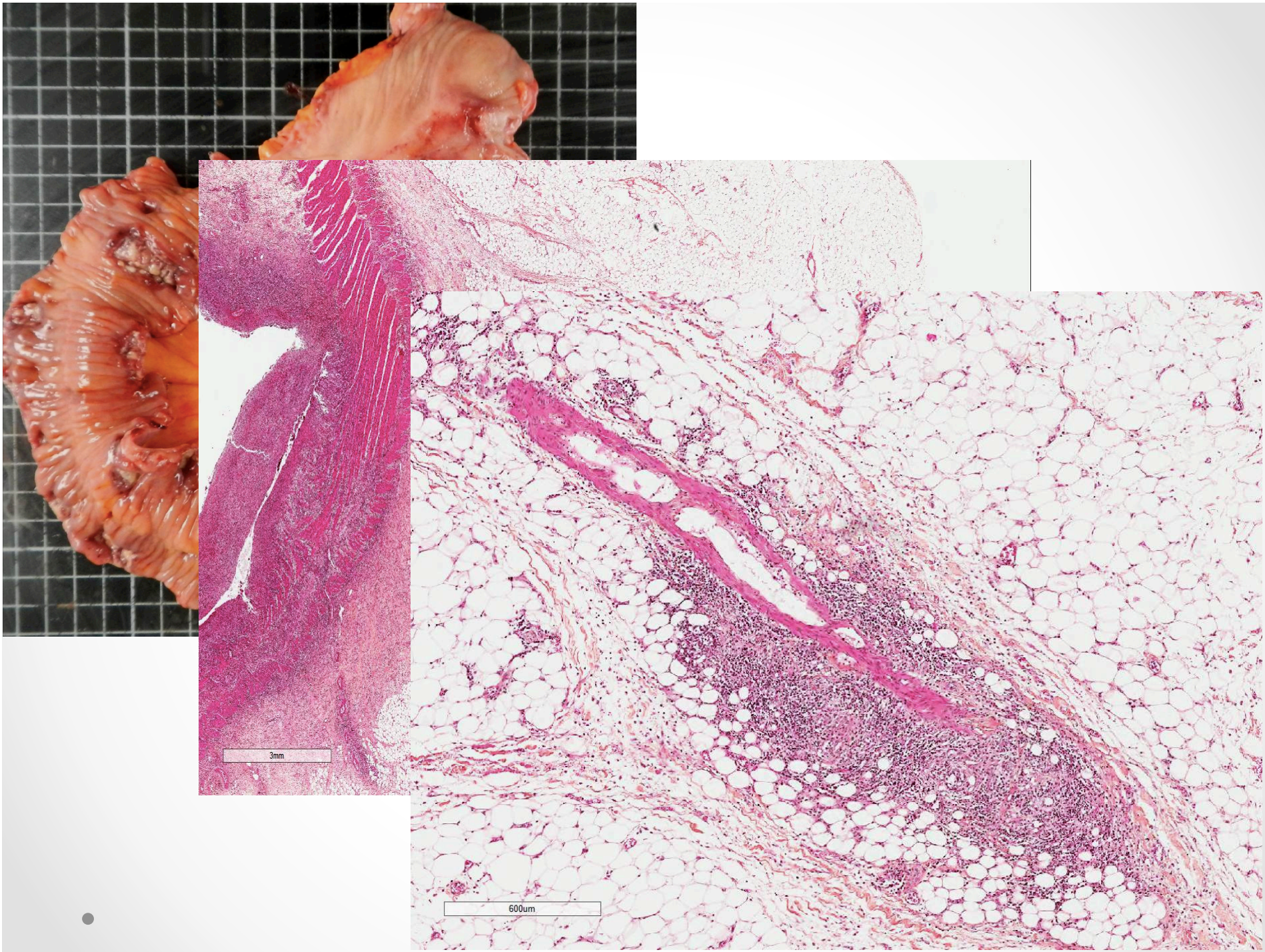


Diagnosis

- Histology of resected piece – vasculitis with predominance of venous vasculitis suggestive of ***Behcet disease***
- Impossible to differentiate Crohn's disease from entero-Behcet on endoscopy with biopsies
- Diagnostic criteria for Behcet disease in this patient NOT MET until histology

Means	*Intestinal BD*	*Crohn's disease*	*Ulcerative colitis*
Endoscopic			
Aphthoid ulcer	+++	-/+	+
Deep round or oval ulcers	+++	0	+
Long swallow ulcers	+	+	+
Skip area	++	-/+	0
Radiologic			
Well-defined deep ulcer	+++	0	0
Skip area	+++	++	0
Fistula formation	++	++	0
Diffuse involvement	0	+	+++
Histologic			
Granuloma	0	+++	0
Vasculitis, venulitis	+++	+	0
Localization			
Ileocecal valv	+++	+++	+/-
Rectum	+/-	+/-	+++
Recurrence after surgery	+++	+++	0
Free perforation	+	+/-	+
Malignancy	0	+/-	+++

Note: 0 = never; +/- = rare; + = occasional: ++ = frequent;
+++ = characteristic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Treatment?

- Multiple choices
  - Cyclophosphamide
  - TNF alpha antagonists
  - DMARDS
- Started on Infliximab 5mg/kg with good evolution

Instead of conclusions

Take-home messages

- Crohn's disease - a systemic disease
- New complications are being described in literature as we speak
- Multidisciplinary approach – gastroenterologists, rheumathologists, dermatologists, ophtalmologists, pathologists and surgeons is the professional way

Thank you!