# **Grand Round Presentation**

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#### Case Report:

-A 51 y/o causasian male was referred to a gastroenterology office by his primary care provider for the evaluation of new onset ascites.

-The patient had been in his basal level of health till about three months prior to presentation at which time he had been prescribed some antibiotic for presumed prostatitis. -A few days after starting the antibiotic he started experiencing diffuse abdominal cramps associated with nausea, vomiting and watery diarrhea.

-He also noticed some staining of the toilet paper with blood but denied any bleeding into the toilet bowl or bloody bowel movements. At this time he also had a low-grade fever.

- -He denied any chest pains or shortness of breath.
- -At about the same time however he also started noticing abdominal swelling which kept on increasing and he presented to his primary care provider and was subsequently referred.

#### -PMH

Hypertension and DJD and spinal disc disease for which he had had knee and cervical spine surgeries in the remote past.

#### - Personal Hx

did admit to heavy alcohol consumption in his younger days a habit he had quit some 25 years ago.

#### Physical Exam

- -middle aged w/m of stocky built, tattoo on each forearm.
- -Vital signs were stable and pt was anicteric
- -He had moderate ascites but none of the other stigmata of chronic liver disease.
- -Rest of the examination was unremarkable.

# LABS

**CBC** 

WBC-16.7,

-Neutrophils-38.9 %,

- Lymphocytes-15%,

HGB-15.3

Platelets -307

ESR-18

BMP Normal Values

#### Liver profile

Alk PO4 -89
AST -19
ALT -14
TP -7.6
Alb -3.8
PT -11

Viral Hepatitis panel - Negative

USN -Liver and GB with dopplers revealed cholelithiasis, shrunken liver, ascites and right pleural effusion.

Stool Studies - Positive for only heavy growth of yeasts, otherwise normal with normal flora

- α1AT, Ceruloplasmin, αFP - normal ANA /ASMA /AMA- Neg

#### CT abdomen and pelvis

- -Marked amount of ascites through the abdomen and pelvis
- -Moderate right and very small left pleural effusions
- -Liver/Spleen/Pancreas nomal
- -Ill-defined increased density in the gall bladder probably representing a stone

CT guided paracentesis
2700 cc of slightly cloudy dark yellow fluid.

-<u>Cells</u>

WBC -8850

Neutrophils -98 %

-100% of these eosinophils

RBC -1900

#### **Chemistry**

TP - 4.5

Albumin - 2.9

**SAAG** - 0.9

Amylase - <30

#### **Microbiology**

Cultures Negative at 72 hours Acid Fast Bacilli (AFB) stain -Negative AFB Culture-Negative at one week

#### -EGD and colonoscopy

- > Gastritis-patchy and scattered throughout the stomach
- > Edematous mucosa throughout the colon except the rectum
  - > Patchy inflammation of the cecum

Biopsies and histopathologic exam revealed mild to moderate eosinophilic inflammation of the stomach, terminal ileum and colon. A diagnosis of Eosinophilic Gastroenteritis was made.

- started on prednisone 40 mg qd.
- made quick recovery of his symptoms, diarrhea resolved as did his ascites and steroid was quickly tapered.
- He was instructed to return in case of need.

One year later the patient returned again c/o mid abdominal pain and being unable to eat because of discomfort. He had recently fallen and hurt his tailbone and had been prescribed a Cox 2 inhibitor.

#### Labs were ordered

#### **CBC**

**WBC** 

- 13.2

N - 47 %, L -20.6%, E - 24.6%

**HGB** 

- 16.3

**Platelets** 

- 233

**ESR** 

- 25

Serum Chemistry - normal Liver panel- Normal CT scan abdomen
Abnormal only for prominent small
bowel loops, gall bladder stone and small
amount of ascites

The patient was restarted on prednisone and made a remarkable turnabout with resolution of his symptoms

- uncommon disease
- first described by Kaijser in 1937 and since then there have been about 300 reported cases.
- true incidence is unknown probably because of varied presentation and lack of diagnostic consideration
- Macroscopically normal areas may have microscopic disease.

- It is characterized by eosinophilic infiltrates of the gut in the absence of parasitic infection.
- Peripheral eosinophilia is common but may be lacking in as much as 20% of cases.
- It affects males and females with equal frequency.

- can affect any layer of the gut.
- depending on whether it affects predominantly the mucosa, muscle layer or serosa or all three layers, it can present as malabsorption syndrome, mimic irritable bowel syndrome (IBS), with obstructive symptoms or even with biliary symptoms.

- -The pathogenesis of EG remains speculative
- Atopy has been implicated

- common association of EG and allergic diseases such as rhinitis, asthma and eczema in as much as 50% of patients
- some pts also have elevated serum IgE levels.
- -food allergy in 50% of patients with mucosal disease
- However this association remains to be proven.

- pathognomonic feature of EG is eosinophilic infiltration of the affected areas of the gastrointestinal tract.
- Once recruited eosinophils perpetuate inflammation by releasing various cytokines such as granulocyte macrophage colony stimulating factor (GM CSF), Interleukin-3 (II 3) interleukin-5 and leukotriene D4 (LTD4) which recruit more eosinophils

- -There have been case reports of drug induced EG. In fact our patient may very well have drug induced EG as suggested by his presentation after taking a certain antibiotic and then recurrence following another medication.
- Could only be proven by rechallenging

		<u>Symptom</u>		
Stomach	43%	Abdominal pain	77%	
Ileum	33%	Anorexia, Nausea,		
Jejunum	31%	Vomiting	48%	
Duodenum	19%	Diarrhea	42%	
Colon & Rectum	11%	Weight Loss	17%	
Diffuse serosal	7%	Abdominal		
		distension	12%	
Others	11%	GI blood loss		
(Gall bladder, Liver,		(occult/overt)	18%	
Esophagus, Spleen, etc)		Ascites	5%	
		Protein malabsorption	Protein malabsorption 9%	

#### Esophageal involvement

- Dysphagia
- Reflux symptoms that is unresponsive to treatment
- Case reports of achalasia occurring in association with eosinophilic esophagitis

#### Small bowel

- Classical manifestations of malabsorption
- It may also present as protein losing enteropathy
- May lead to short stature and failure to thrive in children.

#### Small bowel

Other manifestations of small bowel EG

- diarrhea/ constipation
- features of small bowel obstruction reminiscent of ileiocolic crohns disease
- occult or overt blood loss.

- -There have been reports of EG presenting with colitis and laboratory and ERCP picture of PSC.
- Colon biopsy revealed increased eosinophils and treatment with steroids led to resolution of colitis, liver enzyme abnormalities and ERCP findings

Ascites occurs only with subserosal involvement. However subserosal involvement may also present with any of the features more commonly seen with mucosal or muscular involvement.

- Intraepithelial eosinophils

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- A chronic cytokine driven inflammatory disorder of the esophagus.

- Intraepithelial eosinophils
- A chronic cytokine driven inflammatory disorder of the esophagus.
- Initially described in association with EG, is now clear that in most cases it affects the esophagus in isolation without any involvement of the stomach or the small intestine

- males aged 20-40 years
- most common presenting symptoms are dysphagia and treatment refractory GERD

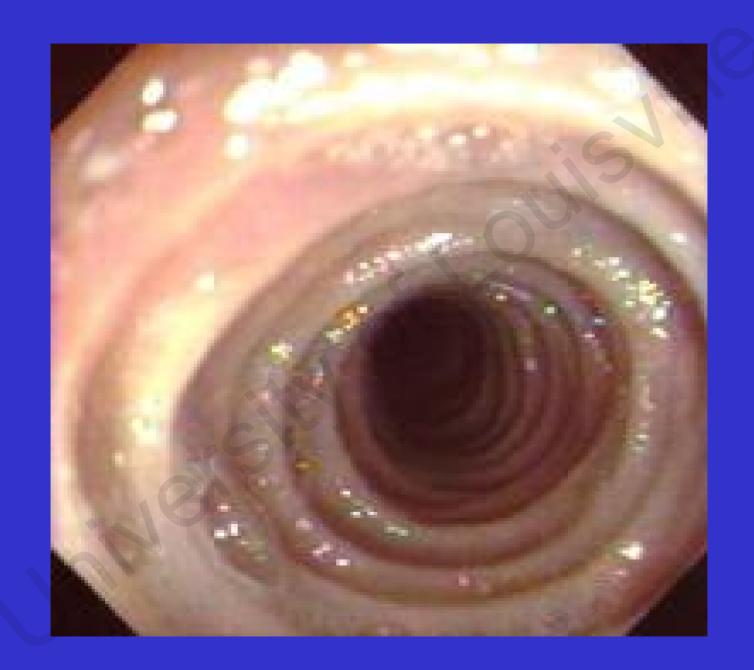
- clinical, histologic & immunopathogenic characteristics of EE has been found to be remarkably similar to the features seen in the tracheobronchial tree of patients with bronchila asthma
- esophagus is thought to undergo remodeling in response to chronic inflammation

- <u>Structurally</u> reflected in the endoscopic appearance of the esophagus
- proximal strictures, rings, corrugation, eosinophilic abscesses and polyps
- esophagus becomes rigid and is more prone to tearing on attempted dilatation
- Functionally- Motility disturbances.



Multiple Esophageal Rings (MER)

- endoscopically the esophagus appears to have concentric rings separated by 1 to 3 mm throughout the length of the esophagus.
- do not flatten out on air insufflation.



Multiple Esophageal Rings (MER)

- histologically characterized by increased eosinophilic infiltration.
- MER was thought to be a consequence of GERD. In one retrospective study done on 24 patients with MER 24-hour pH study was done in 7 patients and failed to a clear-cut evidence of reflux

# Natural History

- 30 patients with histologically proven EE was followed for up to 11.5 years
- recurrent attacks of dysphagia were the commonest presentation
- did not lead to nutritional deficiency or significant deterioration in the quality of life.

# Natural History

- -No increased predisposition to malignancy was noted
- At the time of follow up (longest interval 11.5 yrs) none of the patients were receiving corticosteroids, antihistamines or mast cell stabilizers
- One third of the cohort needed esophageal dilatation -the majority just once.

# **Diagnosis**

-radiological studies are not diagnostic -changes are seen most commonly in the stomach where prominent folds are seen mostly in the antrum. Small bowel and colonic studies similarly show thickened folds and nodularity

# **Diagnosis**

- Confirmed by the characteristic eosinophilic infiltrates on biopsy
- One study of EG used the criteria of 20 eosinophils per high power field as the cut off for defining eosinophilia
- in cases where the disease is purely muscular, endoscopic biopsy may be non diagnostic and may require lapararoscopic full thickness biopsy.

#### **Diagnosis**

- Patients with subserosal involvement present with ascites and the ascitic fluid analysis confirms the diagnosis.

- parasitic infestations
- malignancies
- inflammatory bowel diseases
- hypereosinophilic syndromes
- drugs (ASA, antibiotics, carbamazepine)
- connective tissue disease
- lymphoma, vasculitis (Churg-Strauss syndrome, polyarteritis nodosa)
- celiac disease

#### Natural History

- -Limited experience
- -One study-10 year follow up
- -Spontaneous remission <=> to
- Progression to malabsorption
- -Majority waxing and waning

- -Limited experience
- -No controlled trials

- -Mild Disease = Symptomatic
- antimotility agents
- Iron replacement

- -Severe disease
- -Options
  - -Steroid
  - -Steroid Sparing agents

- -Steroids
  - -most experience
  - -Start at 40 mg/day
  - -maintain till resolution of symptoms
  - -maintenance doses 5-10 mg

- -Steroid sparing agents
  - -limited experiences
  - -case reports
  - -Sodium Cromoglycate
  - -Montelukast (Singular)
  - -Budesonide

#### Treatment

-Ongoing trials at NIH with Anti IL-5 antibody