

CLINICAL CHARACTERIZATION OF EOSINOPHILIC ESOPHAGITIS IN THE MALTESE ISLANDS

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Eosinophilic Esophagitis

- Relatively new diagnosis
 - 1975 first case (adult)
 - Description of 10 cases – Kelly 1995
 - Increasing rates of diagnosis and studies
- Definition:
 - Clinicopathological disease
 - Primary disorder of the esophagus; chronic
 - Esophageal and/or upper GI symptoms
 - Intraepithelial eosinophilia in mucosa
 - Absence of GERD; Lack of response to high dose PPI (2007)
 - ***2011 update: PPI-responsive esophageal eosinophilia

2011 update

- EE to EoE
- Inclusion of terms
 - ▣ Chronic
 - ▣ Immune/antigen driven
- Inclusion of new treatments
- Issue of GERD – can cause eosophageal eosinophilia
- PPI-responsive esophagela esoinophilia
- Some patients with EoE have less than 15 eosinophils/HPF

The Study

□ Aim

- To characterize the paediatric EoE population

□ Method

- Endoscopy database reviewed – 132 individuals
- Chart review of individuals meeting diagnostic criteria for EoE
- Inclusion criteria
 - <18 years, endoscopy done Sept09 – October10, EoE appearance on endoscopy (furlowing, nodularity), eosinophilia (>15 per hpf) noted on esophageal biopsies
- Exclusion criteria
 - Other concurrent illness (e.g. 1 patient with Crohn's)

The Study - Results

- 4 individuals identified
 - 3% of endoscopy population
 - Male 3 vs female 1
 - Age range: 1 – 8 years; Mean age 3.75 years
 - Presentation:

Patient	Symptoms	
1	Vomiting	Food refusal
2	Vomiting	Food refusal
3	Vomiting	Persistent throat clearance
4	Vomiting	

- 2 had neuro-disability
- No history of atopy ← studies show concomitant asthma, atopic dermatitis, food allergy

Neurodisability?

□ Our patients

Patient	Gender	Symptoms	Neurodisability
1	Male	Food refusal	Cerebral palsy
3	Male	Throat clearance	Developmental delay

□ Assad et al

- 89 patients with EoE
- Described relationship to
 - Developmental delay (12%)
 - Seizures (6%)
 - Cerebral palsy (4%)
 - Autism (1%)

□ Pentiuik et al

- described developmental delay in 10/15 children with EoE
- Population of an Interdisciplinary feeding clinic

Post-study....

- Prospective follow up
 - ▣ Treatment with
 - Intensified acid suppression
 - Strict exclusion diet
 - Ingested fluticasone

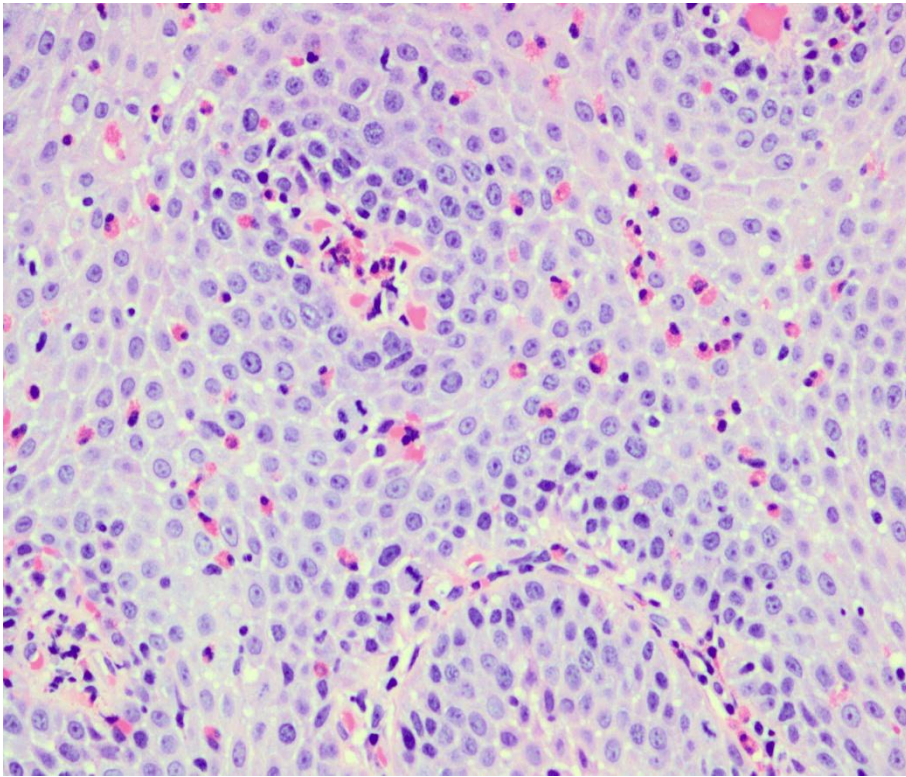
 - ▣ After 1.5 months
 - 3/4 = marked improvement (2 had resolution of symptoms)
 - 1/4 = poor compliance to diet, with persistence of symptoms
 - *All patients underwent repeat endoscopy*

Endoscopy biopsies

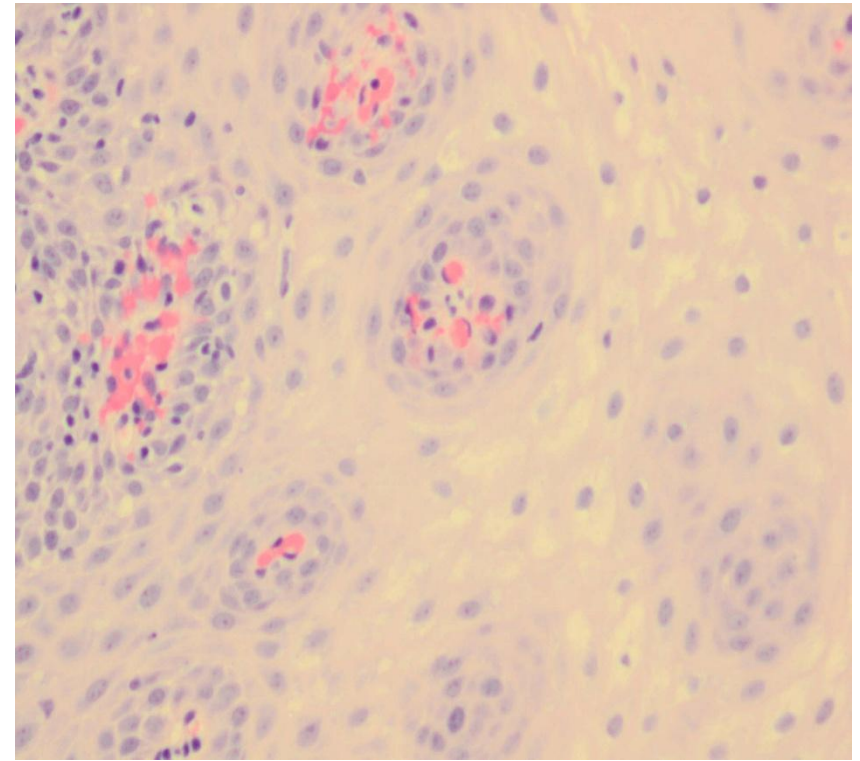
Patient	Biopsy – 1 st endoscopy	Biopsy - Repeat endoscopy
1	“marked infiltration of eosinophils....numbers far exceed 20 per high power field”	“multiple biopsies from oesophagus fail to reveal the presence of any eosinophils”
2	“hyperplastic mucosa...presence of large number of intraepithelial eosinophils”	“upper oesophagus....no eosinophils identified”
3	“biopsies taken from proximal oesophagus, there are large numbers of eosinophils, numbering greater than 40 per high power field”	“practically all high power fields show more than 20 eosinophils”
4	“eosinophils....count in excess of 20/HPF”	“unremarkable oesophageal squamous mucosa”

Histology

□ Patient 1



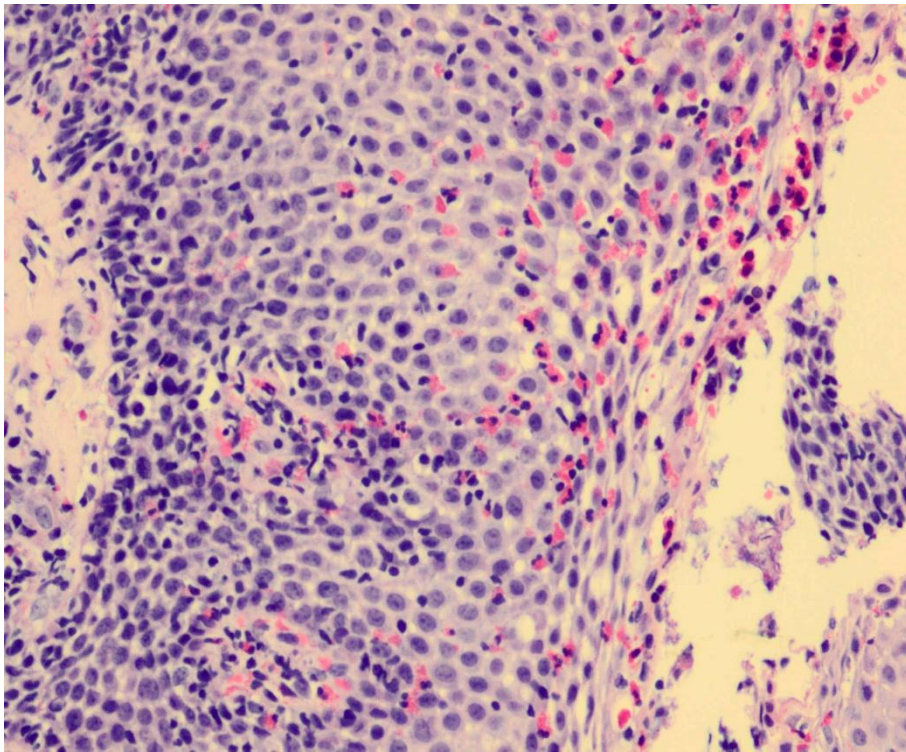
□ Pre-treatment



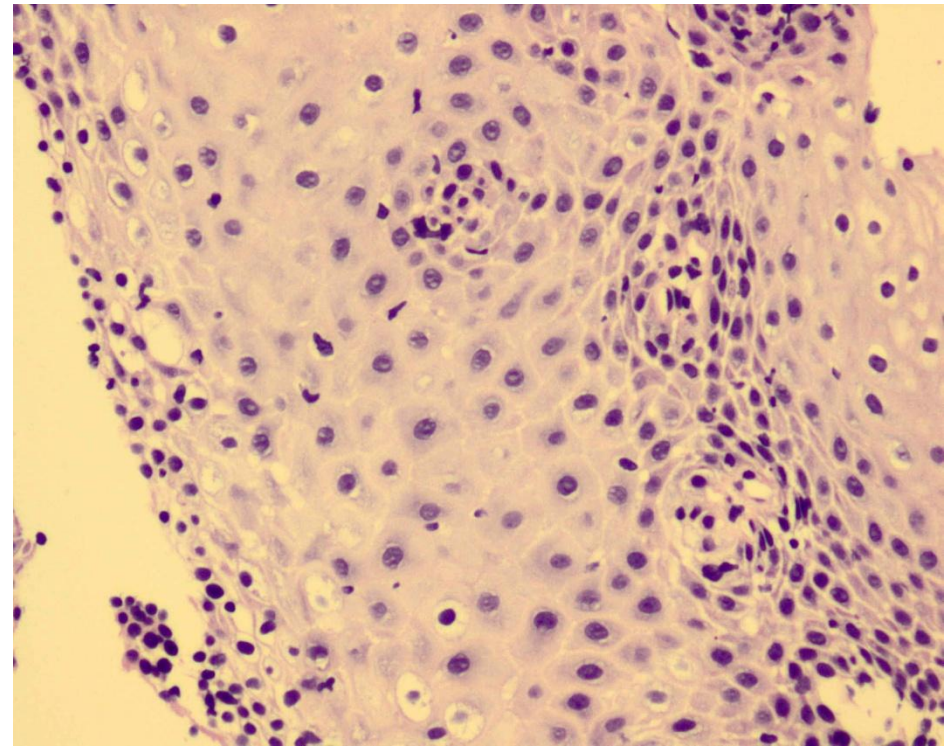
□ Post-treatment

Histology

□ Patient 2



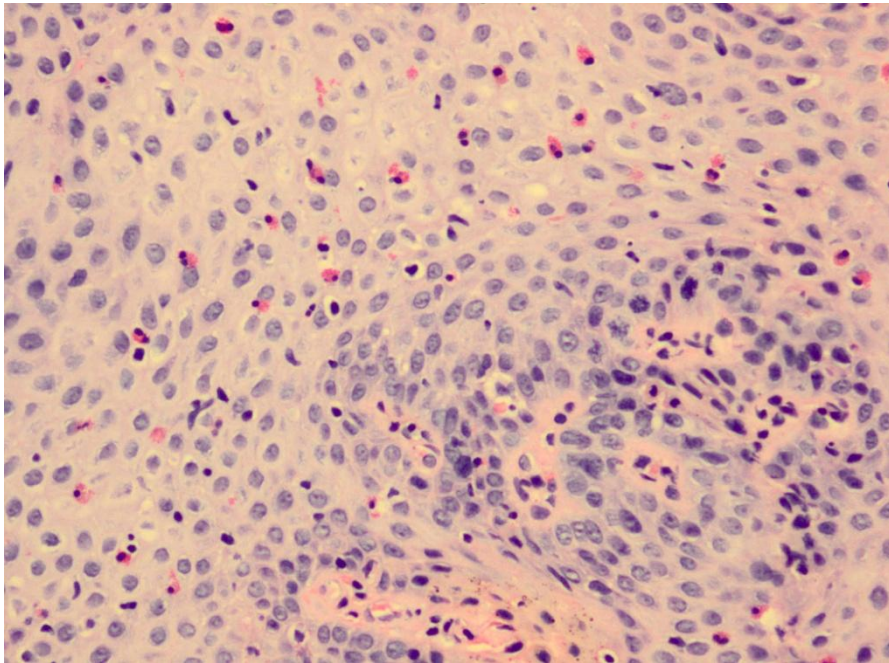
□ Pre-treatment



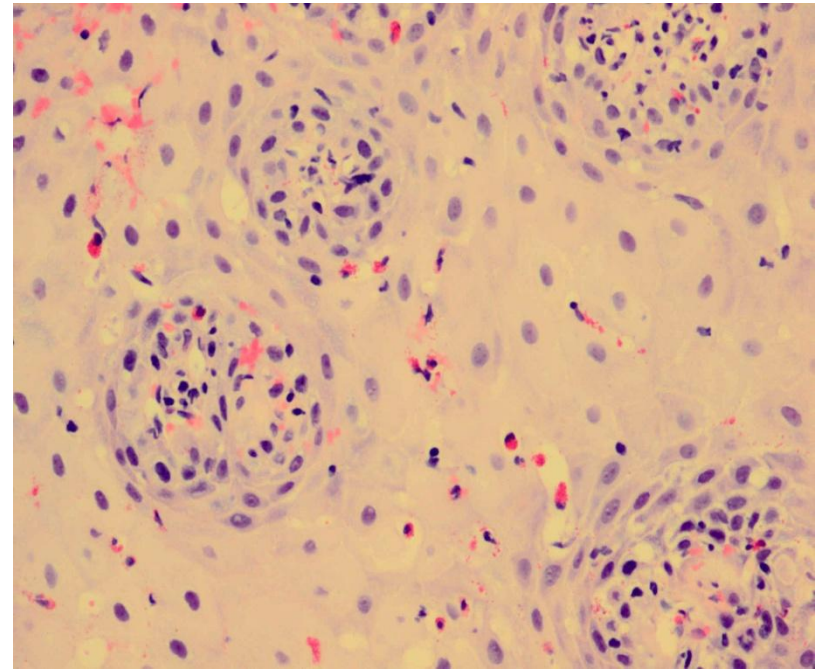
□ Post-treatment

Histology

□ Patient 3



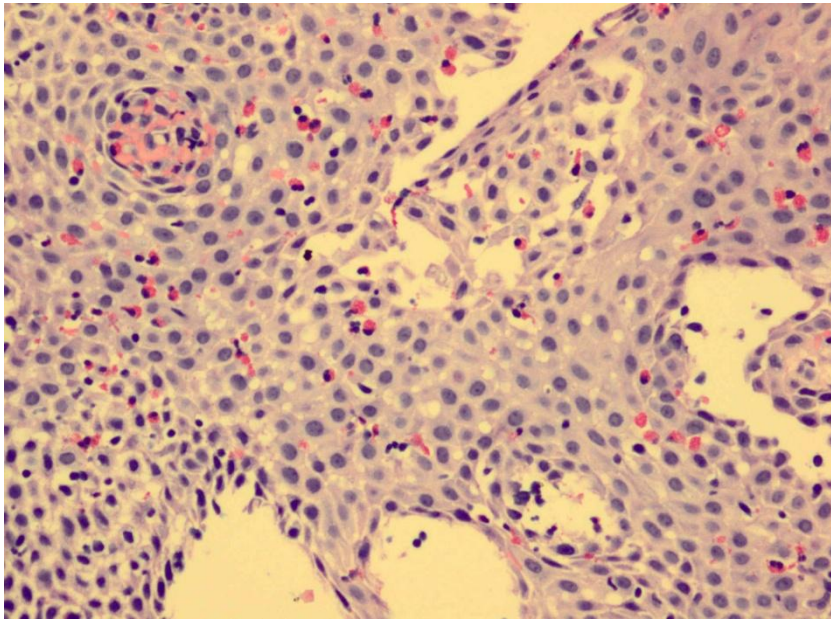
□ Pre-treatment



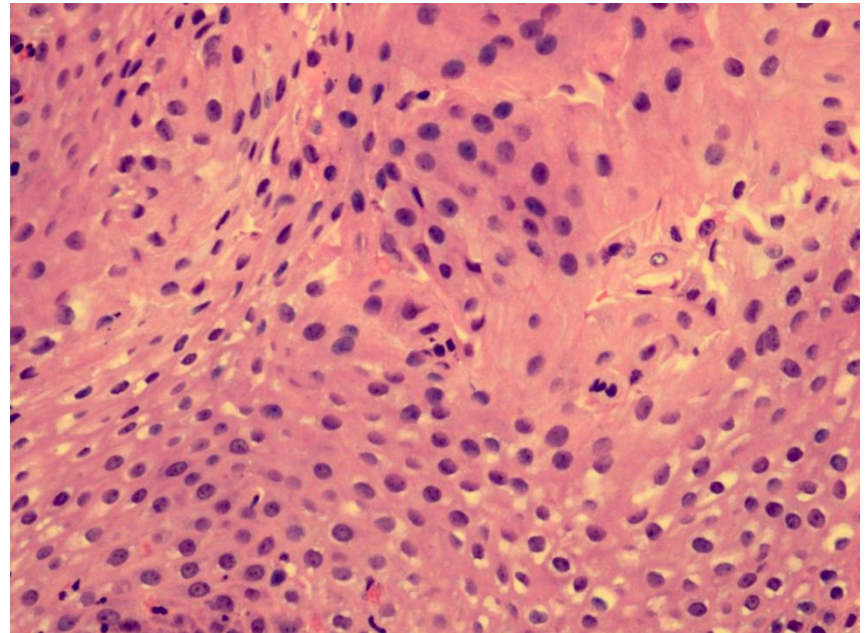
□ Post-treatment

Histology

□ Patient 4



□ Pre-treatment



□ Post-treatment

Recently...

- New patient identified
- 13 year old girl
 - ▣ Presented with dysphagia
- Sister of patient 3
- Biopsy
 - ▣ “intramucosal eosinophils with counts in excess of 100/HPF” in lower oesophagus

- Genetic studies
 - ▣ Candidate gene approach: Eotaxin-3 gene
 - ▣ Genome-wide analysis: EoE susceptibility locus 5q22

Clinical Expression

Clinical expression

- ❑ Typically Caucasian, Male
- ❑ GERD-like symptoms: heartburn and regurgitation
 - ❑ ?Poor response to PPI
- ❑ Dysphagia (adolescents +)
- ❑ Food refusal/ intolerance (infants)
- ❑ Emesis
- ❑ Abdominal pain
- ❑ Failure to thrive

Associations

- ❑ Vary widely
- ❑ Atopic conditions
- ❑ Genetic diseases (e.g. 2 cases Rubenstein-Taybi)
- ❑ Autoimmune disease
- ❑ Coeliac disease
- ❑ Subglottic stenosis
- ❑ Occupational exposure
- ❑ Neurodisability

Work-up

- Endoscopy
 - ▣ Only reliable diagnostic test
 - ▣ No pathognomonic feature
 - ▣ 2 to 4 biopsies, proximal and distal
 - ▣ Described:
 - Longitudinal furrowing (epithelial oedema)
 - White exudates
 - Concentric rings
 - Friability, crepe paper mucosa
 - Biopsy
 - At least 15 eosinophils in one high power field

Treatment

- Diet
 - Restriction
 - Elemental diet
- Pharmacology
 - Steroids
 - Fluticasone – inhaled, topical
 - Montelukast
 - Acid suppression with PPIs
 - Biologics – still being studied

Future challenges

- Still a new diagnosis for our island
- Neurodisability and EoE
- Delay in diagnosis
 - ▣ Overlap with GERD
 - ▣ Earlier start of PPI trial, earlier endoscopy, ?earlier diagnosis

References

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Thanks

