In the name of God
the compassionate
the merciful
Case Report

- At Spring (pollen season), a 29 y/o man; professional singer
- Hx of 9m disphagia, 3m disphonia & globus sensation → ENT clinic
- Hx of asthma, atopy, seasonal rhinitis
- 3 EGDscopy during previous 8y for schatzki ring
- Stroboscopy: Bilat. TVC nodules & post. glottic edema w/o erythema
- EGDscopy & contrast esophagogram: NL → Plan: dilation
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Eosinophilic Esophagitis

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  Otolaryngologic Clinics of North America
  February 2006, 39(1): 205-221

• *Eosinophilic Esophagitis – A mimic of GERD*
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Definition

• a chronic inflammatory disorder of esophagus
• Isolated dense eosinophilic epithelial infiltration
• Unusual structural alteration
• Epithelial proliferative changes

\textit{NOT related to Acid exposure}

History

• 1977 : first description of disease [Dobbins]
• 1978 : labeled as Eosinophilic Esophagitis [Landres]
• 1993 : first comprehensive evaluation in adults [Attwood]
• 1995 : approach to eosinophil in esophagus was changed
• 2002 : first case report of EE in ENT journals
Epidemiology

• Annual incidence: 1 / 10,000
  (GERD incidence in ENT practice: 4-10%; in Pediatrics: 18%)
• Prevalence in patients with refractory reflux symptoms: 68 – 94%
• more reported in children; in adults are common in 2nd to 4th decade
• male dominance (M/F = 3/1)
• May have a genetic predisposition

Differential diagnoses of Eosinophilic infiltration in Esophagus

- Gastroesophageal reflux disease
- Eosinophilic Esophagitis
- Infections (parasitic, fungal)
- Peripheral hypereosinophilic synd.
- Allergic gastroenteropathy
- Inflammatory bowel disease
- Esophageal leiomyomatosis
- Hodgkin’s disease
- Periarteritis
- Scleroderma
- Drug injury
- Recurrent vomiting
Etiology / Pathophysiology

- Occurs due to Allergic and Immunologic responses

- Operative immune responses against Ag:
  - IgE mediated (extrinsic & allergic) → skin prick test or RAST are Positive
  - non IgE mediated (intrinsic & nonallergic) → tests are Negative

- Eosinophil: tissue / blood ratio = 100-300 / 1
  (majority in GI but Very few in esophagus and some argue that must be 0)

- IL-5 plays a central role in eosinophilic infiltration

- Possible contributors:
  - Food allergens
  - Environmental Aeroallergens

Relationship between GERD and EE  → No Causality
Clinical Manifestations

Common features:
- GERD symptoms refractory to antireflux (including fundoplication)
- Intermittent Dysphagia with or without food impaction
- Peripheral blood eosinophilia [31%]
- Increased IgE levels [55%]

Symptoms in children:
- vomiting
- regurgitation
- epigastric pain
- poor eating (food refusal or dysphagia)
- failure to thrive
- hematemesis
- waterbrash

Most children have atopic conditions (asthma, dermatitis, food allergy [milk, soy])

Any child who presents with food impaction that requires endoscopic extraction should be evaluated for the possibility of EE.
Clinical Manifestations

Symptoms in adults:

- Dysphagia [93%] (often with impaction [62%])
  - Many required endoscopic dilation (even misdiagnosed with Schatzki)
  - Usually longstanding in duration (date back to childhood)
  - Not associated with significant wt loss & Compensation with eating slowly
- Allergic Hx with lower prevalence than pediatrics are present [52%]
- GERD symptoms (Heartburn [52%], vomiting, odynophagia, noncardiac chest pain)
- Mean duration of symptoms until Dx: 7y (max. reported: 15y)

Upper airway & respiratory symptoms & signs:

- rhinosinusitis
- cough
- wheezing
- pneumonia
- globus
- hoarseness / dysphonia
- subglottic stenosis
- chronic laryngeal edema

Presenting symptoms: usually GI in nature
Diagnosis

• **Endoscopy with Biopsy is required to Dx.**

• **Endoscopic finding : quite subtle to very obvious**
  - NL endoscopy [9%]
  - Subtle esophageal mucosal granularity
  - Adherent white exudates [16%]
  - Concentric rings (trachealization) [49%]
  - Vertical linear furrowing
  - Loss of NL vascular pattern (edema)
  - Fragile mucosa (crepe – paper appearance [after endoscopy]) [59%]
  - Esophageal mucosal tearing almost uniformly after dilation
  - Focal esophageal stenosis (64% in proximal or mid esophagus)
  - Long segment strictures [5%]

Nonerosive lesions in full length of esophagus

• **Multiple biopsies from different levels of esophagus are necessary**
  - Eosinophilic infiltration are not homogenous
  - Dense infiltration may be present with normal appearance
Diagnosis

- **Pathologic findings:**
  - Nonspecific findings of epithelial inflammation
    - basal cell hyperplasia & increased papillary size
    - *is seen in both EE & GERD*
  - Quantity of eosinophilic infiltrated *(differentiates EE & GERD)*
    - cut-off value is controversial
    - most accepted: eosinophil $\geq 20$ in HPF ($\times 400$)
    - in one study, eosinophil $\geq 7$ in HPF provides:
      - 85% accuracy for diagnosing EE
      - 86% predictive value for failure with antireflux therapy
  - Histology not changed with antireflux therapy

- **Definitive Dx:** reflux like symptoms resistant to antireflux therapy
  + NL or borderline pH metry + isolated eosinophilic infiltration
  (response to steroid in important for diagnosis)
Treatment

• **Food elimination**
  - an elemental diet with amino-acid based formula (not palatable)
  - it’s effectiveness was demonstrated in pediatrics
  - has not been studied extensively in adults

• **Medical treatment**
  ➢ **Corticosteroids**
    ➢ **topical**: the most effective with the least side effects [95%]
      - Fluticasone Spray (220 µg per puff) / 4 puffs / BD after meal / 6 weeks
        - Teach the patient a “Bad inhaler technique”
        - For decreasing dry mouth: rinse the mouth with water
        - For candidiasis: Nystatin($10^5$U/5mL) swish & swallow / Daily / 5 days
          - Nasal Fluticasone mixed in chocolate syrup
    ➢ **Systemic**: best reserved for relapses or resistance
      - Proved effectiveness BUT has potential serious side effects
Treatment

• **Medical treatment**
  - Leukotriene receptor antagonist
    - Montelukast (selective eosinophil LT-D4 receptor inhibitor)
      - number of eosinophils does not decreased but symptoms are dissapeared
  - Anti IL-5 monoclonal antibody
    - Mepolizumab: for severe cases / 6 weeks IV / no serious side effects
  - Proton Pump Inhibitor: Symptom relief in 27%
  - Cromolyn sodium

• **Endoscopy & Dilation**
  - adjunctive treatment [immediate improvement in 83%]
  - traditional mechanical dilation is more effective than ballon dilators
  - symptomatic recurrence after dilation: 3 – 8 months
  - risk of nucosal tearing is high but perforation is rare [1.5%]
    (severe chest pain after dilation that needed to admission in 10%)

• **Criteria for response**: symptom improvement + decrease in infiltration
Prognosis

- Symptom-free intervals after Rx withdrawal is patient-dependent
- Eosinophilic inflammation is confined to esophagus
- No potential for malignancy or hypereosinophilic Syndrome
- Quality of life indicators show:
  - Socioprofessional activities is affected moderately in 3% & mildly in 50%
  - Serious food impaction is experienced in 71% of adults & 24% of pediatrics
- If disease develops during childhood, often outgrows with patient
- If untreated → fibrosis and stenosis are possible
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- EGDscopy & Bx from proximal, mid, & distal of esophagus
- 41 eosinophil / HPF → Dx: Eosinophilic Esophagitis
- Plan: 6w oral fluticasone & PPI → Improve in all symptoms & signs
Case Report 2

- A 2 y/o girl with intolerance to many food products, choking, vomiting
- Past Hx of prematurity & 3w intubation &mild to moderate stridor after extubation
- With Dx of subglottic stenosis underwent multiple laser procedures
- Endoscopy: grade III SGS, hypopharyngeal & postcricoid inflammation
- Double-probe pH monitoring & Esophagoscopy: NL
- Esophageal Bx: 10 – 12 Eosinophil/HPF
- Plan: antireflux & cricotracheal resection & intubation for 6d
- SGS recurred & did not respond to serial dilation
- Postoperation was complicated by difficulty with oral intake (retching & vomiting)
- Allergy test: NL CBC, elevated IgE, positive Skin Test
- Underwent laryngotracheal reconstruction with costal cartilage graft
- Postoperation developed thigh breathing & intermittent stridor
- Anaphylactic reaction to peanut → emergent intubation & medical Rx
- Dexamethasone 0.5 mg/Kg → clinical & endoscopic improve but Persisted grade II SGS
- After 6w → side effects of steroid → discontinuing steroid → side effects disappear
- Return of symptoms → aerosolized steroid & strict diet → markedly improve within 2w
Disease is of antiquity and nothing about it changes. It is we who change as we learn to recognize what was formerly imperceptible.

Jean Martin Charcot
(1825 – 1893)