Diseases of the esophagus

Manar Hajeer, MD, FRCPath
University of Jordan, School of medicine
A hollow, highly distensible muscular tube

Extends from the epiglottis to the GEJ, located just above the diaphragm
Diseases that affect the esophagus

1. Obstruction: mechanical or functional.
3. Inflammation: esophagitis.
4. Tumours.
Mechanical Obstruction

- Congenital or acquired.

- Examples:
  - Atresia
  - Fistulas
  - Duplications
  - Agenesis (v rare)
  - Stenosis.
Atresia

- Thin, noncanalized cord replaces a segment of esophagus.
- Most common location: at or near the tracheal bifurcation
- +/- fistula (upper or lower esophageal pouches to a bronchus or trachea).
Clinical presentation:

- Shortly after birth: regurgitation during feeding
- Needs prompt surgical correction (rejoin).

Complications if w/ fistula:

- Aspiration
- Suffocation
- Pneumonia
- Severe fluid and electrolyte imbalances.
Esophageal stenosis

- Acquired >> Congenital.
- Fibrous thickening of the submucosa & atrophy of the muscularis propria.
- Due to inflammation and scarring

Causes:
- Chronic GERD.
- Irradiation
- Ingestion of caustic agents
Clinical presentation

- Progressive dysphagia
- Difficulty eating solids that progresses to problems with liquids.
Efficient delivery of food and fluids to the stomach requires coordinated waves of peristaltic contractions.

**Esophageal dysmotility**: discoordinated peristalsis or spasm of the muscularis.

**Achalasia**: the most important cause.
Achalasia

- **Triad:**
  - Incomplete LES relaxation
  - Increased LES tone
  - Esophageal aperistalsis.

- Primary >>>secondary.
- Pneumatic balloon dilatation of the LES

- Lower esophageal sphincter
Primary achalasia

- Failure of distal esophageal inhibitory neurons.
- Idiopathic
- Most common
Secondary achalasia

- Degenerative changes in neural innervation

- Intrinsic
- Vagus nerve
- Dorsal motor nucleus of vagus

- Chagas disease, Trypanosoma cruzi infection >> destruction of the myenteric plexus >> failure of LES relaxation >> esophageal dilatation.
Clinical presentation

- Difficulty in swallowing
- Regurgitation
- Sometimes chest pain.
Achalasia-like disease

- Diabetic autonomic neuropathy
- Infiltrative disorders (malignancy, amyloidosis, or sarcoidosis)
- Dorsal motor nuclei lesions (produced by polio or surgical ablation).
Vascular diseases: Esophageal Varices

- Tortuous dilated veins within the submucosa of the distal esophagus and proximal stomach.
- Diagnosis by: endoscopy or angiography.
ESOPHAGEAL VARICES

- Esophagus
- Stomach
Dilated varices beneath intact

Robbins Basic Pathology 10th edition
Pathogenesis:

- **Portal circulation**: blood from GIT >> portal vein >> liver (detoxification) >> inferior vena cava.
- Diseases that impede portal blood flow >> portal hypertension >> esophageal varices.
- Distal esophagus: site of Porto-systemic anastomosis.
- **Portal hypertension**: collateral channels in distal esophagus >> shunt of blood from portal to systemic circulation >> dilated collaterals in distal esophagus >> varices
Portal system

Usual circulation

Portal circulation

SITES OF PORTACAVAL ANASTOMOSIS

Five sites of portal/systemic circulation:
1. Lower third of the Esophagus
2. Paraumbilical Area
3. Upper end of Anal canal
4. Retroperitontial
5. Bare area of liver

https://www.slideshare.net/charslan626/hepatic-anastomosis
Causes of portal hypertension

- Cirrhosis is most common
  - Alcoholic liver disease.
- Hepatic schistosomiasis 2nd most common worldwide.
Clinical Features

- Often asymptomatic.
- Rupture leads to **massive hematemesis and death**.
- 50% of patients die from the first bleed despite interventions.
- Death due to: hemorrhage, hepatic coma, and hypovolemic shock
- Rebleeding in 20%.
ESOPHAGITIS

- Esophageal Lacerations.
- Mucosal Injury
- Infections
- Reflux Esophagitis
- Eosinophilic Esophagitis
Esophageal Lacerations

- **Mallory weiss tears are most common**
- Due to: severe retching or prolonged vomiting
- Present with hematemesis.
- Failure of gastroesophageal musculature to relax prior to antiperistaltic contraction associated w/ vomiting >> stretching >>> tear.
Linear lacerations
- longitudinally oriented
- Cross the GEJ.
- Superficial
- Heal quickly, no surgical intervention
Chemical Esophagitis

- Damage to esophageal mucosa by irritants
- Alcohol,
- Corrosive acids or alkalis
- Excessively hot fluids
- Heavy smoking
- Medicinal pills (doxycycline and bisphosphonates)
- Iatrogenic (chemotx, radiotx, GVHD)
Clinical symptoms & morphology

- Ulceration and acute inflammation.
- Only self-limited pain, odynophagia (pain with swallowing).
- Hemorrhage, stricture, or perforation in severe cases
Infectious esophagitis

- Mostly in debilitated or immunosuppressed.
- Viral (HSV, CMV)
- Fungal (candida >>> mucormycosis & aspergillosis)
- Bacterial: 10%.
Candidiasis:

- Adherent.
- Gray-white pseudomembranes
- Composed of matted fungal hyphae and inflammatory cells
Esophageal Candidiasis

white-yellowish patchy distribution

https://www.pinterest.com/pin/374291419013418659/
www.researchgate.net/publication/285369734_Esophageal_Candidiasis_as_the_Initial_Manifestation_of_Acute_Myeloid_Leukemia
Herpes viruses
Punched-out ulcers
Histopathologic:
Nuclear viral inclusions
Degenerating epithelial cells ulcer edge
Multinucleated epithelial cells.
Figure 4: Gastroendoscopic findings revealed the presence of multiple...
CMV:
- Shallower ulcerations.
- Biopsy: nuclear and cytoplasmic inclusions in capillary endothelium and stromal cells.
Reflux Esophagitis

- Reflux of gastric contents into the lower esophagus
- Most frequent cause of esophagitis
- Most common complaint by patients
- Gastroesophageal reflux disease, GERD

- Squamous epithelium is sensitive to acids
- **Protective forces**: mucin and bicarbonate, high LES tone
Pathogenesis

- Decreased lower esophageal sphincter tone
  (alcohol, tobacco, CNS depressants)
- Increase abdominal pressure
  (obesity, pregnancy, hiatal hernia, delayed gastric emptying, and increased gastric volume)
- Idiopathic!!
MORPHOLOGY

- **Macroscopy (endoscopy)**
  - Depends on severity (Unremarkable, Simple hyperemia (red))

- **Microscopic:**
  - Eosinophils infiltration
  - Followed by neutrophils (more severe).
  - Basal zone hyperplasia
  - Elongation of lamina propria papillae
Clinical Features

- Most common over 40 years.
- May occur in infants and children
- Heartburn, dysphagia,
- Regurgitation of sour-tasting gastric contents
- Rarely: Severe chest pain, mistaken for heart disease
- Tx: proton pump inhibitors
Complications

- Esophageal ulceration
- Hematemesis
- Melena
- Strictures
- Barrett esophagus (precursor of Ca.)
Eosinophilic Esophagitis

- Chronic immune mediated disorder
- **Symptoms:**
  - Food impaction and dysphagia in adults
  - Feeding intolerance or GERD-like symptoms in children
- **Endoscopy:**
  - Rings in the upper and mid esophagus.
- **Microscopic:**
  - Numerous eosinophils w/n epithelium
  - Far from the GEJ.
Most patients are: atopic (atopic dermatitis, allergic rhinitis, asthma) or modest peripheral eosinophilia.

Tx:
- Dietary restrictions (cow milk and soy products)
- Topical or systemic corticosteroids.
- Refractory to PPIs.
Barrett Esophagus

- Complication of chronic GERD
- Intestinal metaplasia within the esophageal squamous mucosa.
- 10% of individuals with symptomatic GERD
- Males >> females, 40-60 yrs
- Direct precursor of esophageal adenocarcinoma
- Metaplasia >> 0.2-1% /year >> dysplasia >> adenocarcinoma.
MORPHOLOGY

- **Endoscopy:**
  - Red tongues extending upward from the GEJ.

- **Histology:**
  - Gastric or intestinal metaplasia
  - Presence of goblet cells
  - +/-Dysplasia: low-grade or high-grade
  - Intramucosal carcinoma: invasion into the lamina propria.
Normal squamous  Barrett’s oesophagus  Barrett’s oesophagus with low-grade dysplasia  Barrett’s oesophagus with high-grade dysplasia  Adenocarcinoma

Population screening  Predicting prognosis, best therapy and response

Predicting risk of progression and response to preventive therapy

Baishideng Publishing Group
Management of Barrett

- Periodic surveillance endoscopy with biopsy to screen for dysplasia.
- High grade dysplasia & intramucosal carcinoma needs interventions.
ESOPHAGEAL TUMORS

- Squamous cell carcinoma (most common worldwide)
- Adenocarcinoma (on the rise, half of cases)
Adenocarcinoma

- Background of Barrett esophagus and long-standing GERD.
- Risk factors: dysplasia associated Barrett, smoking, obesity, radioTx.
- Male : female (7:1)
- Geographic & racial variation (developed countries)
Pathogenesis

- From Barrett >> dysplasia >> adenocarcinoma
- Acquisition of genetic and epigenetic changes.
- Chromosomal abnormalities and TP53 mutation.
MORPHOLOGY

- Distal third.
- Early: flat or raised patches
- Later: exophytic infiltrative masses

- Microscopy:
- Forms glands and mucin.
Clinical Features

- Pain or difficulty swallowing
- Progressive weight loss
- Chest pain
- Vomiting.
- Advanced stage at diagnosis: 5-year survival <25%.
- Early stage: 5-year survival 80%
Squamous Cell Carcinoma

- Male : female (4:1)
- Underdeveloped countries.

- **Risk factors:**
  - Alcohol
  - Tobacco use
  - Poverty
  - Caustic injury
  - Achalasia.
  - Plummer-Vinson syndrome
  - Frequent consumption of very hot beverages
  - Previous radiation Tx.
Pathogenesis

- In western: alcohol and tobacco use.
- Other areas: polycyclic hydrocarbons, nitrosamines, fungus-contaminated foods
- HPV infection implemented in high risk regions.
MORPHOLOGY

- Middle third (50% of cases)
- Polypoid, ulcerated, or infiltrative.
- Wall thickening, lumen narrowing
- Invade surrounding structures (bronchi, mediastinum, pericardium, aorta).
Microscopy:

- Pre-invasive: Squamous dysplasia & CIS.
- Well to moderately differentiated invasive SCC.
- Intramural tumor nodules
- Lymph node metastases:
  - Upper 1/3: cervical LNs
  - Middle 1/3: mediastinalparatracheal, and tracheobronchial LNs.
  - Lower 1/3: gastric and celiac LNs.
Clinical Features

- Dysphagia
- Odynophagia
- Obstruction
- Weight loss and debilitation
- Impaired nutrition & tumor associated cachexia
- Hemorrhage and sepsis if ulcerated.
- Aspiration via a tracheoesophageal fistula
- Dismal Px: 5 year survival <9%