

# **Clinical Case Conference**

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# Chief complaint: neck swelling, cough

49 F with h/o

- intramural esophageal pseudodiverticulosis
- esophageal perforation 2/2 food impaction in 04 s/p partial esophagectomy c/b esophageal stricture s/p multiple dilations
- multinodular goiter, hyperthyroid
  
- recent adm 2 mos. ago for dysphagia & FTT, had J-tube placed c/b intractable vomiting, TPN initiated

## 49F w neck swelling and cough

- eval in endocrine clinic for abnl TFTs and noted to have ant. neck swelling/pain, SOB x 3 days, sent in to ED
- c/o worsening anterior neck pain, productive cough, fatigue & weakness

# Clinical Case

## PMH/PSH:

- colonic diverticulosis
- depression
- anemia
- allergic rhinitis

## Allergies:

- PCN, Aspirin

FH: NC

## Medications:

- Prevacid qd, zofran, tylenol prn
- SSRI, flonase

## Social History:

- etoh: quit 7yrs ago, prev abuse
- Tob: quit 7yrs ago, prior 2ppd for 15yrs
- never illicit

# Physical Examination

Vitals: T38.5, HR 125, 104/60, 22, 99% RA

- **GEN:** chronically ill-appearing, nontoxic
- **HEENT:** no scleral icterus, **PERRLA**, **OP** clear
- **Neck:** +anterior neck swollen, nontender, no crepitus, no mobile mass palpable
- **CV:** tachy, regular, no mrg
- **Lungs:** bibasilar crackles, diffuse rhonchi, +coughing clear secretions
- **Abdomen:** **NABS**, soft, **NT**, **ND**
- **Ext:** warm, no **LEE**
- **Skin:** no pallor

# Labs

<b>140</b>	<b>109</b>	<b>5</b>	<b>&lt; 101</b>
<b>3.5</b>	<b>25</b>	<b>0.7</b>	

<b>28.1</b>	<b>8.5</b>	<b>&lt; 308</b>
<b>26</b>		

**N: 85%**

**INR 1.4, PT 15.8, PTT 27.7**

## A bit more history re: dysphagia

- as of 5 mos ago, swallowing good without anything getting stuck. Occ chest burning which occurs related to certain foods and only situational
- last EGD 5 mos ago w narrowing
- CT surgery continued to dilate esophagus
- over last 2 months, progressively worse dysphagia to solids
- currently on TPN for nutrition, J-tube for meds, water po daily however comes right back up recently

# Prior EGDs

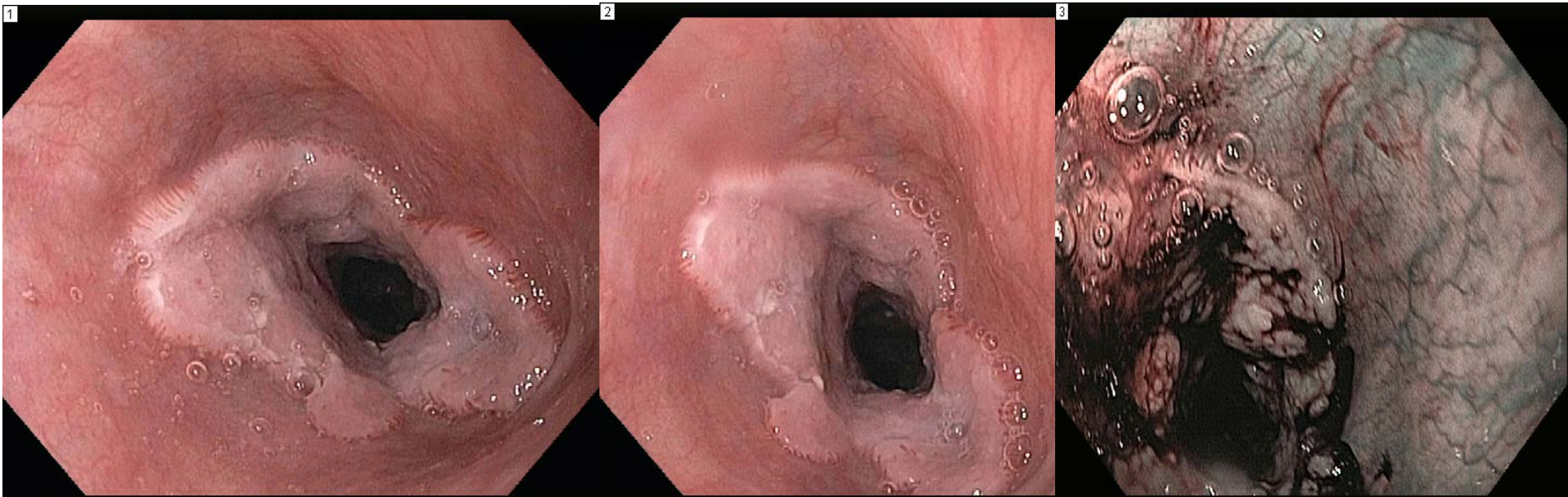
**EGD 7/04:** Starting at 20cm, area of extensive heterotopic gastric mucosa, distal to this was diffusely nodular mucosa with tiny punctate openings, exudate covering in parts and stricture formation but permitting passage of scope, extending to distal esophagus but with last 5cm of esoph appearing nl. Area of prior perf w sinus tract visualized.

Bx: Reactive non-keratinizing squamous epithelium, no dysplasia

**EGD 3/11:** Circumferential white, raised lesion w vascular margins at 20cm. Scope does not pass. Difficult to manage airway given proximal lesion.

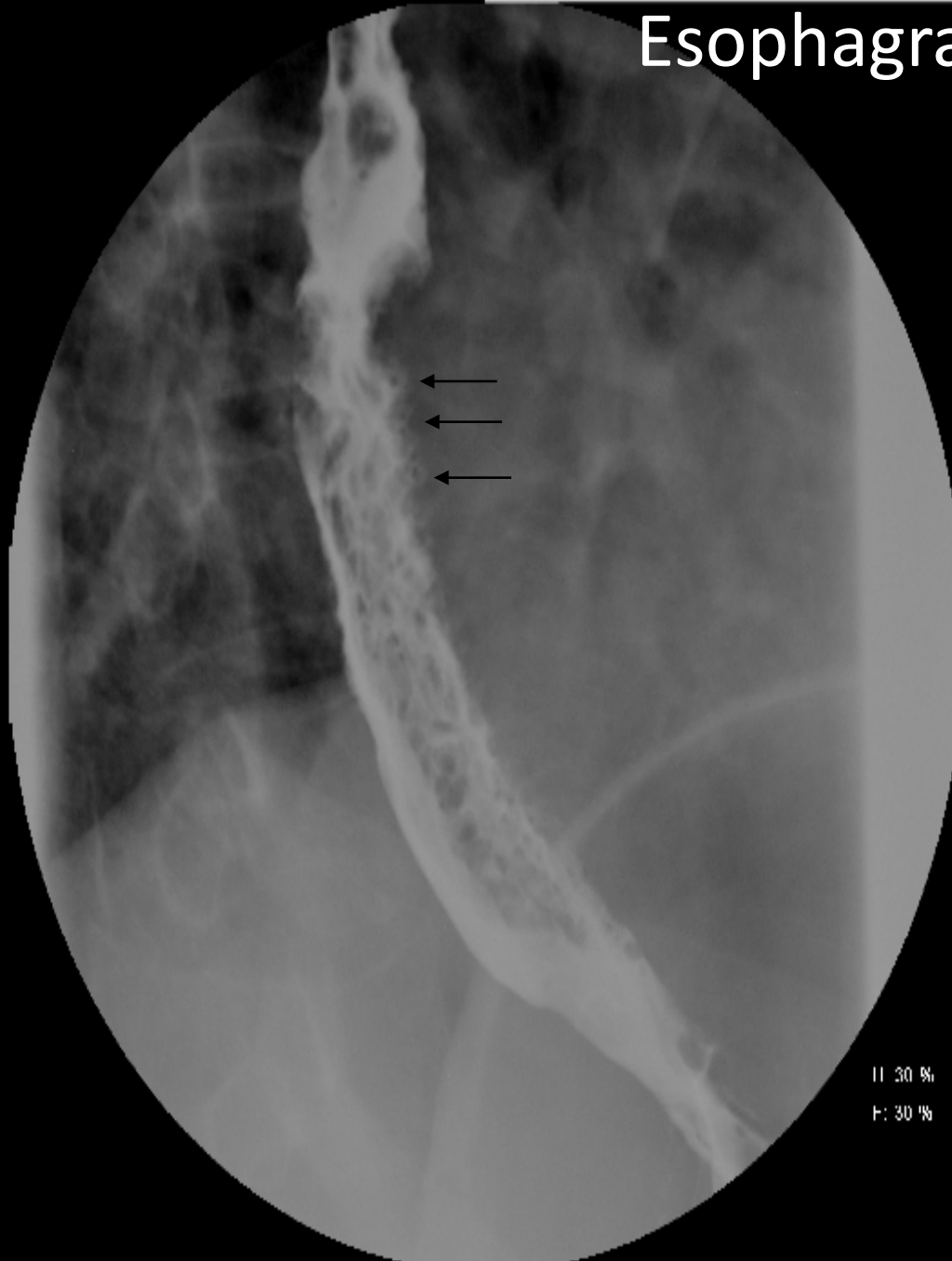
Bx: Squamous mucosa w glycogen acanthosis

Cytology: reactive squamous cells in background of mild acute and chronic inflamm. No carcinoma.



Se:21  
Im:1

# Esophagram



H: 30 %

F: 30 %

# Video Esophagram 3/11

-There are **tiny flask-shaped outpouchings from the upper esophagus** just below the thoracic inlet. Stricture at the junction of the cervical and thoracic esophagus on the prior barium study of April 2004 is no longer seen. However, there is a markedly abnormal lower half of the thoracic esophagus, which has a grossly irregular contour associated with what appear to be multiple polypoid defects, especially in the mid and distal esophagus. The etiology of these findings is uncertain.

## CONCLUSION:

1. Esophageal intramural pseudodiverticulosis.
2. Markedly abnormal thoracic esophagus with grossly irregular contour and multiple polypoid defects of uncertain etiology; possible etiologies include Candida esophagitis with fungus balls and plaques versus benign or malignant esophageal neoplasms.

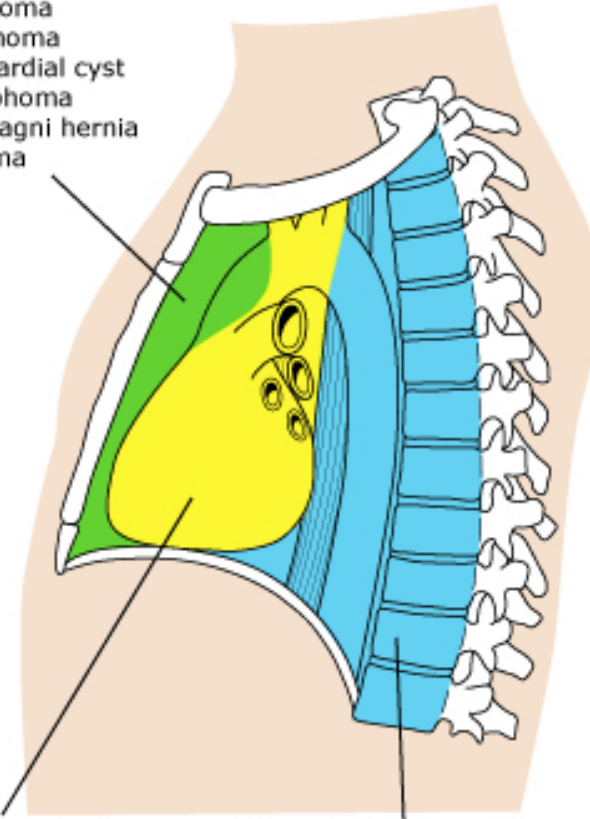
# Questions

- **What is your DDx for this patient's now pw neck swelling, cough, dysphagia?**
- **What diagnostic studies do you want next?**

# Possibilities of Mediastinal Masses

## **Anterosuperior mediastinum**

- Goiter
- Ascending aortic aneurysm
- Parathyroid tumor
- Esophageal tumor**
- Angiomatous tumor
- Teratoma
- Thymoma
- Pericardial cyst
- Lymphoma
- Morgagni hernia
- Lipoma



## **Middle mediastinum**

- Lymphoma
- Lymph node hyperplasia
- Bronchogenic tumor
- Bronchogenic cyst

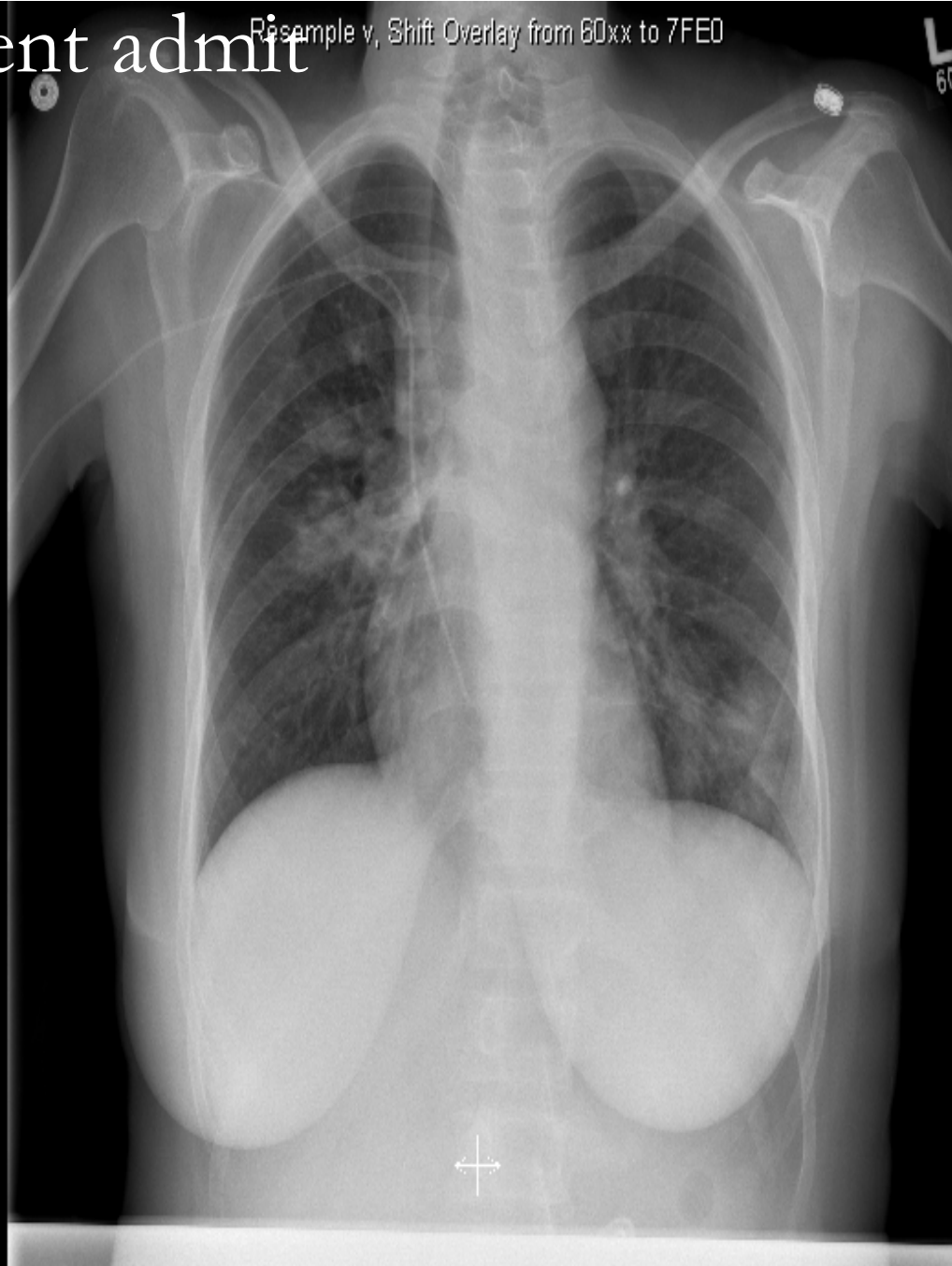
## **Posterior mediastinum**

- Neurogenic tumor
- Aortic aneurysm
- Enteric cyst
- Hiatal hernia
- Esophageal tumor**
- Bronchogenic tumor
- Paraspinal abscess
- Extramedullary hematopoiesis

# On current admit

Resample v, Shift Overlay from 60xx to 7FE0

Se:2  
Im:1



L  
60

W Chest PA

C1817  
W3614

## **CXR:**

1. Bilateral patchy infiltrates in the right upper lobe and left lower lobe likely due to aspiration given history.
2. Deviation of the trachea to the right

**Neck Ultrasound** showed nl thyroid but large mass displacing trachea



Se:5  
Im:26

[A]

[R]

[L]



[P]

Thorax 5.0 B31s  
ISOVUE 370

C-600  
W1465

## CT CHEST noncontrast

1. **Markedly thickened esophagus with multiple leaks**, extending into the mediastinum, left main bronchus, right bronchus intermedius, and right lower lobe.
2. **Multifocal patchy opacities** involving the lungs bilaterally
3. Mild mediastinal and prob hilar lymphadenopathy

## CT NECK w contrast

1. Masslike thickening of the thoracic esophagus in region of prior anastomosis. **Assoc loculated collections about this mass** c/f possible necrotic lymph nodes vs perforations/abscess. Dilatation of lower cervical esophagus **with assoc laryngeal mass-effect**
2. Patchy peribronchial opacities in R hemithorax c/f aspiration vs infectious PNA given mass effect from esophageal mass.

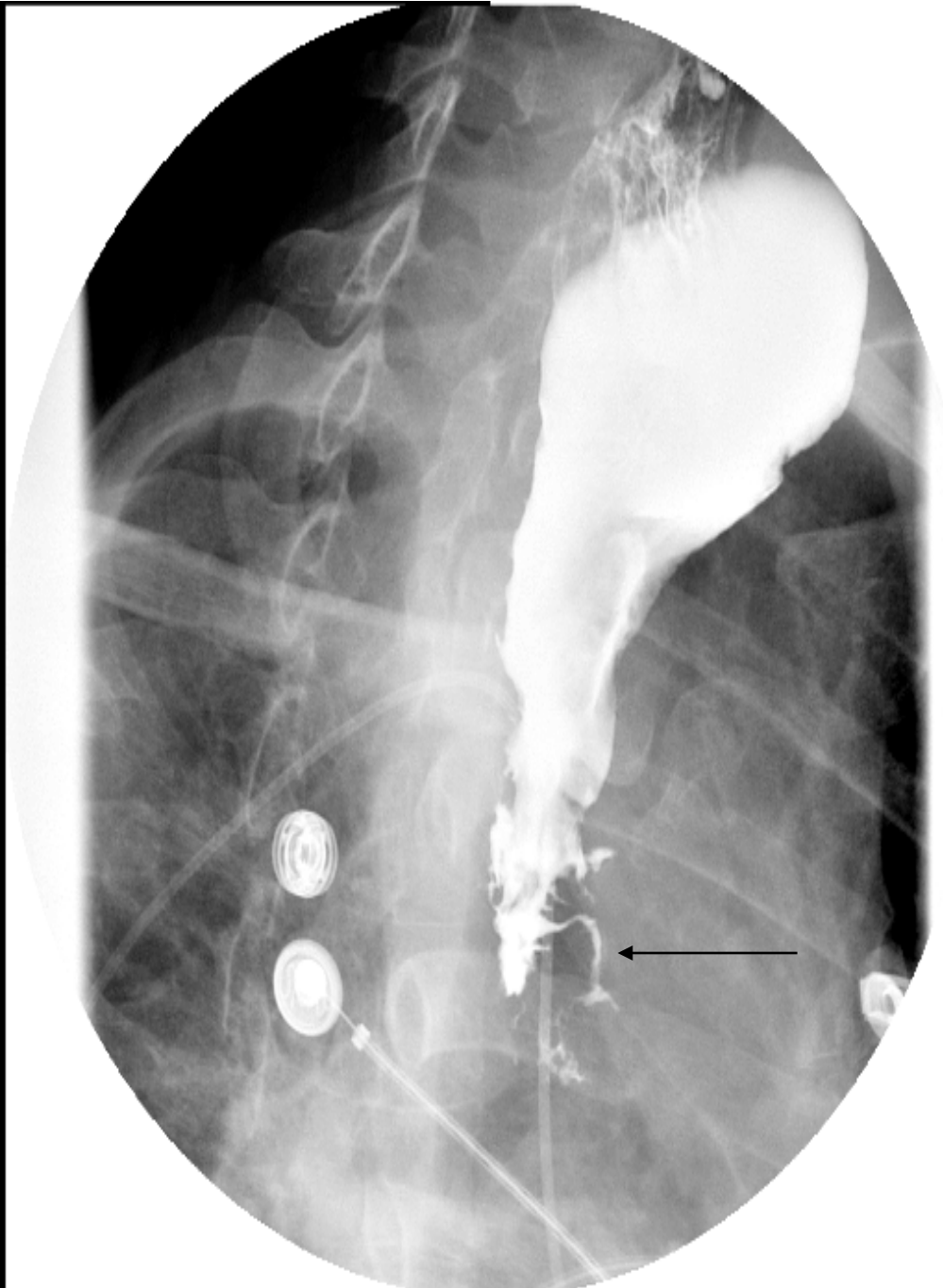
**What would you like to do next?**

A) UGI series

B) EGD

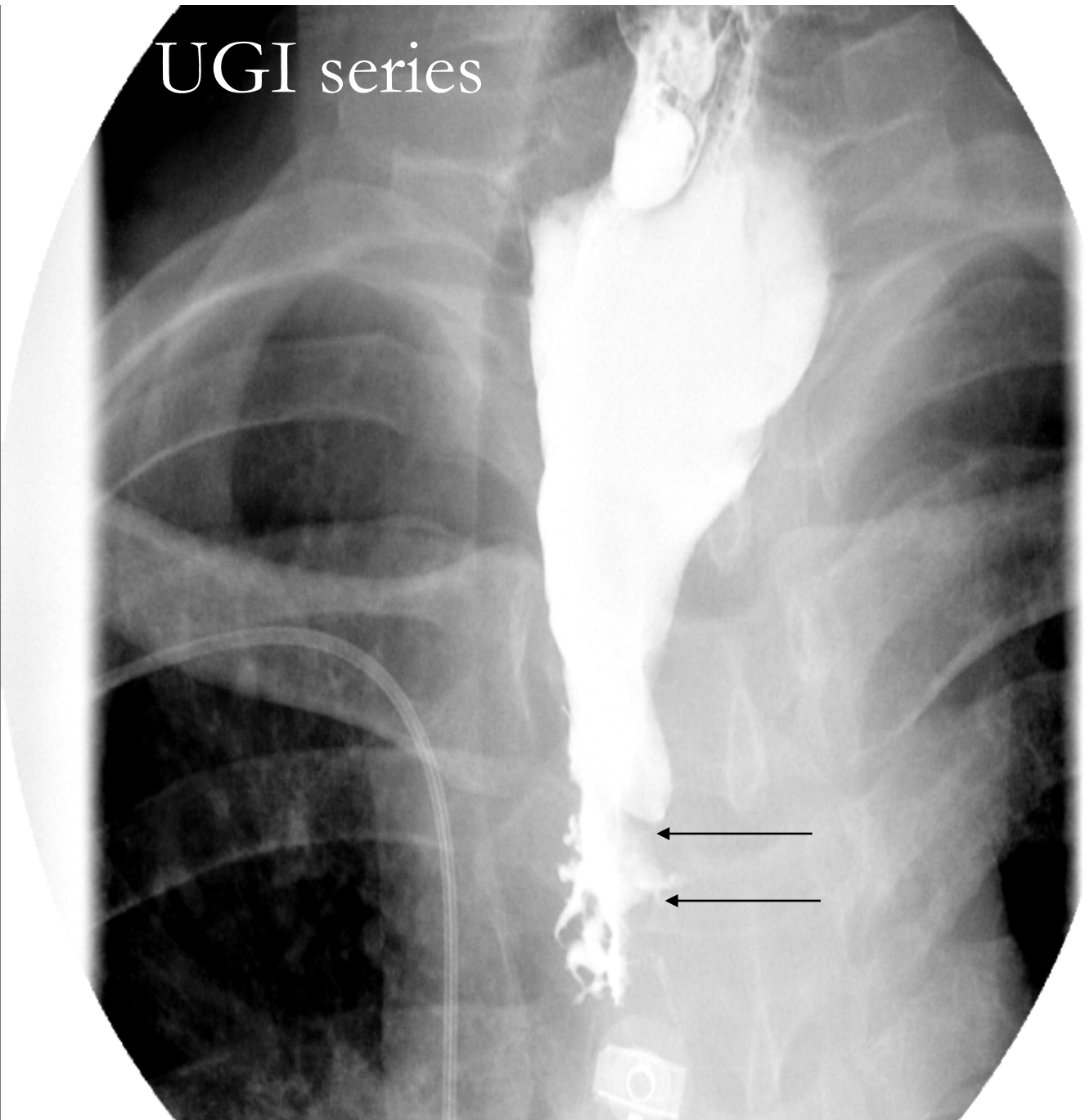
C) Bronchoscopy

D) Trt PNA and no further work-up



Se:7  
Im:1

# UGI series



02 01

## **UGI series:**

Complete obstruction of upper to mid thoracic esophagus with multiple small, contained tracks/collections presumably representing tiny sealed-off leaks in this region. A developing esophagobronchial fistula at site of obstruction, seen to enter the left main bronchus

## Hospital course

-Started on Vanc/cefepime/flagyl

Bronchoscopy by CT surgery

Fungating tumor on posterior wall of L main bronchus  
and medial wall of R main bronchus. Bx taken

Peds+Adult esophagoscope

Esoph inflammed w multiple polypoid protrusions and  
hypertrophied squamous islands

Stricture at 25cm unable to be passed

-Unable to clear secretions, re-intubated → ICU

# Tracheal Tumor Path

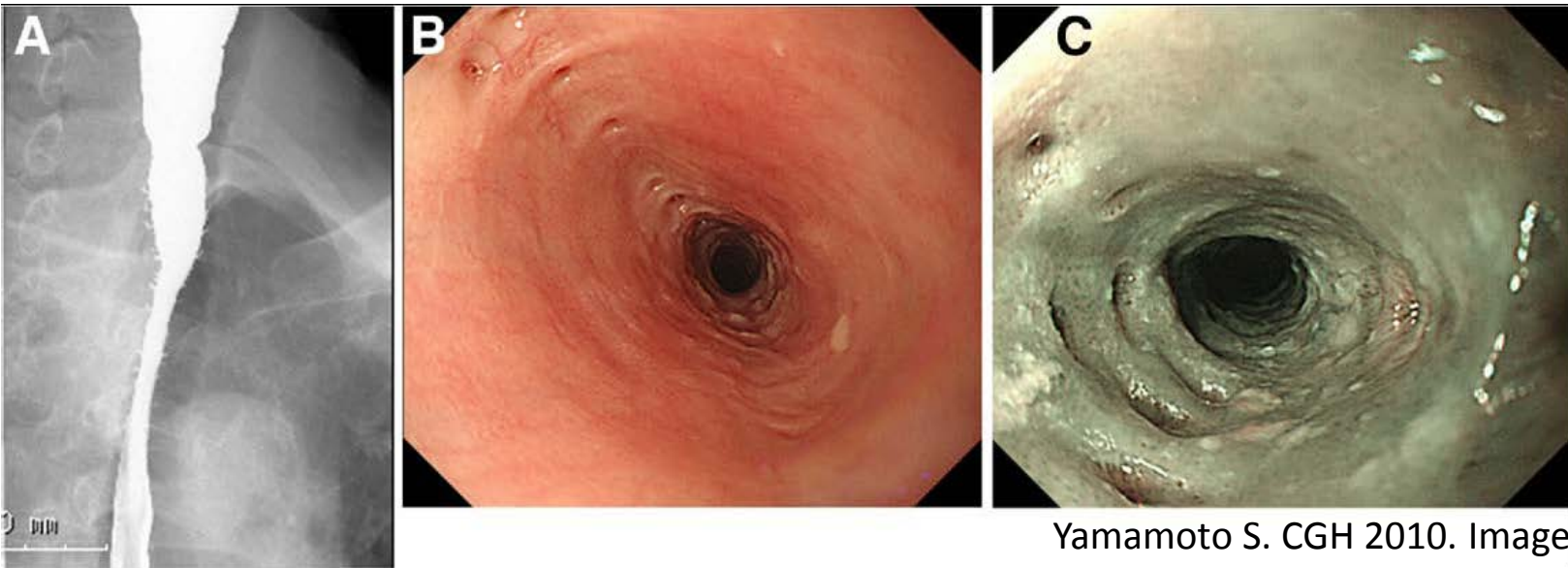
1. Atypical squamous epithelium c/w **invasive well differentiated squamous cell carcinoma**. Necrotic debris with atypical dyskeratotic cells present with inflamed respiratory mucosa.
2. Multiple previous esophageal biopsies noted. A fragment showing markedly hyperplastic squam mucosa with glycogenation and nuclear atypia, most c/w invasive well differentiated squamous cell **carcinoma of the esophagus**, most likely arising in the setting of long-standing achalasia.

# Topics of discussion

- Intramural esoph pseudodiverticulosis
- Esophageal Squamous cell carcinoma
- Malignant tracheoesophageal fistula
- Endoscopic palliation of esoph cancer
- Esophageal stenting

# Intramural esoph pseudodiverticulosis

- **Benign**
- **Multiple flask-shaped out-pouchings of the esophagus**
- **Represent dilated submucosal excretory ducts**
- **Uncommon: <200 cases reported in literature**



Yamamoto S. CGH 2010. Image of month  
PubMed Case reports 1990-2010

# Intramural esoph pseudodiverticulosis

- Assoc *Candida* infection (50%), reflux esophagitis, esophageal malignancy
- Manifest with intermittent solid food dysphagia (stricture formation and/or achalasia)
- Rx: esophageal dilation of sx strictures, trt *Candida*, PPI, bx to r/o malignancy
- ?increased risk of esophageal CA

# Esophageal SCC

- Esophageal neoplasia rarely benign
- ~14,000 new cases in 2003 (13,000 deaths anticip)
- Lifetime risk 0.8% in men, 0.3% in women
- Squamous CA most common
- Higher incidence in African Americans M
- Assoc Tob, etoh
- Other risks: achalasia, celiac, caustic injury, tylosis
- Progressive dysphagia, 5yr survival rate <10%
- Inoperable SCC generally incurable

# TNM stage definitions for esophageal cancer

## Primary tumor (T)\*

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- Tis High-grade dysplasia•
- T1 Tumor invades lamina propria, muscularis mucosae, or submucosa
- T1a Tumor invades lamina propria or muscularis mucosae
- T1b Tumor invades submucosa
- T2 Tumor **invades muscularis propria**
- T3 Tumor invades adventitia
- T4 Tumor invades adjacent structures
- T4a Resectable tumor invading pleura, pericardium, or diaphragm
- T4b **Unresectable tumor** invading other adjacent structures, such as aorta, vertebral body, trachea, etc.
- NX-N3 mets in >7 nodes
- M0, M1 distant mets
- G1-G4 undifferentiated

# Locally advanced, unresectable

- Trachea indentation or fistula
- Distant mets (excluding celiac nodes)
- No preservation of fat planes btwn on esoph and surrounding
- Palliative resection not mainstay
- Chemoradiation (5-FU, cisplatin)+EBRT
  - Cervical: similar to ENT SCC +/- salvage op
- Neoadj chemo/xrt to downstage -> resectable: no improved survival

# Malignant TE fistula

- **Dismal prognosis**
- **Complication of tumor, chemo/XRT, or recurrence**
- **If caused by chemo/XRT, tends to resolve after completion**
- **If persistent, options:**
  - **Cervical esophagostomy, gastrostomy, J-tube**
  - **Stenting: airway, esophageal, or both**
- **XRT contraindicated**
- **Barium study: avoid hyperosmolar agent**

# Endoscopic palliation of esoph CA

- **Esophageal Dilation (repeat)**
- **Laser Therapy (mult sessions)**
- **Injection therapy (etoh, cisplatin/epi)**
- **EMR (if early)**
- **Photodynamic Therapy**
  - **Photosensitiz agent inj, laser exposure-> necrosis**
  - **Skin photosensitivity**
- **APC**
- **Brachytherapy**
- **Endoscopic Stenting (SEPS < SEMS)**

# Esophageal stenting

- **Goal is palliation: dysphagia, nutrition**
- **Types: uncovered, partially covered, fully covered**
- **Complications**
  - **Migration, Stridor, Reflux/Aspiration**
- **>95% w malignant obstruction tolerate liquids post-SEMS**
  
- **For TE fistula**
  - **Covered**
  - **Outcomes: 70-100% successful closure**
  - **Removability**
  - **Airway stenting prior if tracheal compression**

# Back to our pt

- Extubated, completed 10d antibiotics**
- CT a/p: mult intra-Abd lymphadenopathy**
  
- Not surgical candidate**
- Onc no plans for palliative chemo/XRT in light of persistent, mult fistulae & infection risk**
  
- Esophageal stent considered but wld need too extensive airway stenting deemed high risk of resp compromise by Interv pulm**
  
- Discharged to home hospice**

# Discussion questions

- Mgmt of Intramural esoph pseudodiverticulosis
- Interval surveillance period of above
  - indeterminate path, achalasia, inflamm stricture
- Number of biopsies needed +/- brushings
- Role for chromoendoscopy
- Dilate to preop staging w EUS (celiac axis node, depth of invasion)

# Prior path

1. From EGD 3/11: No evidence of carcinoma. Most fragments reveal hyperplastic squamous mucosa, marked spongiosis, glycogenation and mild focal acute inflammation. Focal area with surface epithelial changes with mild nuclear enlargement but without marked nuclear membrane irregularity is noted. Concurrent esophageal brush sample also reveals only reactive changes. Esophageal biopsy is squamous mucosa with **acute inflammation and focal atypia, favor reactive**.

2. From Bx 7/11:

Esophagus, biopsy:

Squamous mucosa with acute inflammation and focal atypia, **favor reactive**. While the biopsy is favored to be reactive in nature, it is noted that the brushings show atypia that is felt to be beyond what is encountered in reactive states (abnormal shapes with dyskeratosis and marked nuclear atypia).

Cytology Final Interpretation

Atypical and dykeratotic squamous cells, mild acute inflammation.

No AFB seen, few Candida

3. From Bx 8/11:

Esophagus, 27-34 cm, biopsy x4:

**Atypical squamous epithelium admixed with inflammation**

Necrotic debris and inflammation. There is extensive cytologic atypia of the squamous epithelium, numerous dyskeratotic cells, extremely **worrisome for a neoplastic process**. Cannot definitively exclude squamous cell carcinoma.

4. Repeat Bx 8/11:

Esophagus, biopsy x2:

Atypical squamous epithelium with acute inflammation  
Squamous cell carcinoma cannot be excluded.

**Thank you**