May 07, 2013

Cystic Fibrosis Program Committee  
Teaching Cases  
Cystic Fibrosis Foundation  
6931 Arlington Road  
Bethesda, Maryland 20814

RE: Case submission for Physician’s Grand Rounds session

Dear Cystic Fibrosis Program Committee,

I am submitting a case of a 54 year old cystic fibrosis (CF) patient with eosinophilic esophagitis (EoE) leading to gastric outlet obstruction. The patient presented with acute pancreatitis that was treated at an outside hospital and referred to our CF center for follow up. Her serum amylase was 111 and lipase was 1743. In despite of the fact that her amylase and lipase improved, she continued to have epigastric pain and vomiting. Her abdominal plain films and a right upper quadrant ultrasound were unremarkable. CT scan abdomen showed distal esophageal thickening and dilated stomach. Floroscopic esophagogram revealed normal esophagus, gastroparesis and gastric outlet obstruction (GOO). Nuclear gastric emptying study confirmed gastroparesis. She underwent upper gastrointestinal endoscopy which showed multiple furrows in distal 10 cm of the esophagus and pre-pyloric stenosis causing obstruction without mass or ulceration. Biopsies of esophagus and gastric stenosis revealed >40-60 (eos/hpf) confirming EoE and GOO. She underwent serial dilations of gastric stenosis with endoscopy but failed to show improvement. The patient was treated with metoclopramide, proton pump inhibitor (PPI) and oral viscous budesonide (OVB) for EoE. On follow up 3 months later she had complete resolution of nausea, abdominal pain and weight loss.

Goralski et al reported a case series of pediatric CF patients with EoE, and noted that EoE is underappreciated in CF because of symptom overlap with other common gastrointestinal disorders (J. of Cystic Fibrosis 12 (2013) 9-14) This case provides the opportunity for discussion of the importance of diagnosing GOO with EoE in CF. Additionally, it offers opportunity for discussion of the therapeutic options available in managing EoE and its complications in CF. To our knowledge EoE progressing to GOO has not been reported previously. Our case is an example of such presentation. She was treated with medications and showed complete recovery without surgery.

Thank you for your consideration.

Sincerely,

Nauman Chaudary, MD  
Adult Program Director,  
Assistant Professor,
**History and Clinical Findings:**
PS is a fifty four year old female with cystic fibrosis (ΔF508/unknown). She was diagnosed with CF at age 50 after presenting with recurrent sinusitis requiring sinus surgery and sweat chloride of 67. She has normal spirometry and is pancreatic sufficient. She has not required previous hospitalization for pulmonary or gastrointestinal symptoms.

Other past medical history is remarkable for gastro-esophageal reflux (GERD), hysterectomy, iron deficiency and migraine. She has chronic *P. aeruginosa* in her sputum cultures.

In September of 2012, PS presented to an outside hospital with 24 hour history of severe epigastric pain and 1 week history of nausea and vomiting, flatulence and indigestion. She was found to have acute pancreatitis and transferred to our CF center for admission. Vital signs were normal. Physical exam was unremarkable for abdominal tenderness or organomegaly. She was taking proton pump inhibitors (PPI) for gastro-esophageal reflux disease (GERD). Her labs were unremarkable for hepatitis, cholecystitis and urinary infection. Chest xray ruled out pneumonia, abdominal plain film ruled out ileus and a right upper quadrant ultrasound did not show gall stones. She continued to c/o abdominal pain despite the fact that her serum amylase and lipase improved. CT scan abdomen and pelvis revealed distal esophageal thickening and dilated stomach. She was discharged home after 4 days of hospitalization tolerating regular diet and improvement in her nausea and vomiting with outpatient follow up.

At 1 week follow up she continued to have nausea and vomiting with intermittent abdominal pain. Evaluation included floroscopic esophagogram which revealed normal esophagus, gastroparesis and gastric outlet obstruction (GOO). MRI abdomen showed liver cysts. Nuclear gastric emptying study confirmed gastroparesis with 4 hour gastric retention of 37%. She underwent upper gastrointestinal endoscopy (UGI) and colonoscopy (LGI). LGI showed melanosis coli diffusely. UGI showed multiple furrows in distal 10 cm of the esophagus, diffuse food, liquid in stomach and severe pre-pyloric stenosis causing obstruction without mass lesion or ulceration. Biopsies of mid, distal esophagus, gastric and pyloric stenosis revealed >40-60 eos/hpf confirming EoE with infiltration of pre pyloric area. She underwent serial dilations of pyloric stenosis and failed to show improvement. Repeat floroscopic esophagram with small bowel follow through and nuclear gastric emptying study showed persistent GOO and gastroparesis. Exhaled nitric oxide levels (FENO) were at upper limits of normal for age suggesting eosinophilic inflammation. The combined results were felt to be consistent with eosinophilic esophagitis infiltrating the pre-pyloric area causing stenosis and gastric outlet obstruction.

**Follow-up and outcome:**
The patient was started on metoclopramide 10mg po q.i.d, omeprazole 40 mg po b.i.d and oral viscous budesonide slurry (1mg/2ml) b.i.d. At follow up 3 months later she had complete resolution of nausea, abdominal pain and weight loss. Patient tolerated the medications well. She is taking regular diet and successfully managed without surgical correction for her GOO.

**Conclusion:**
EoE is an increasingly recognized cause of epigastric pain, heartburn and nausea that is unresponsive to PPI in CF. The association of CF and EoE is not likely because CF is a genetic disease where as EoE is an allergen mediated disease. Atopic diseases like asthma are more common in people with EoE. Duration of therapy for EoE remains uncertain. It is also not known whether EoE progresses to gastrointestinal malignancy. EoE may commonly cause esophageal stricture and rupture may occur from food impaction. Since FENO in CF is generally <5 ppb, a value at the upper limit of normal could be interpreted in this clinical setting as possibly elevated. To our knowledge EoE progressing to GOO has not been reported previously. Our case is an example of such presentation. She was treated with medications and showed complete recovery without surgery.