Learning Objectives
- Become familiar with typical and atypical presentation of appendiceal tumors
- Be able to identify epidemiology of appendiceal carcinomas
- Recognize current treatment options

Patient Presentation
A 65 year old male with a history of hypertension, type II DM and hypercholesterolemia who presented for outpatient surveillance colonoscopy after a screening colonoscopy at age 50 revealed polyps and had a subsequent follow up colonoscopy that was normal. The patient complained of dark, tarry stools with 70lbs weight loss over four months as well as a change in stool caliber.

Evaluation
Colonoscopy: 15 mm sessile polyp in cecum at appendiceal orifice 10cm polyposid mass at sigmoid colon
Pathology: Tubular adenoma with high grade dysplasia Invasive, mod differentiated cystadenocarcinoma
CEA: 6.23ng/ml (normal)
*Total colectomy with ileo-rectal anastomosis
Stage: pT4 pN1 cM0 2/12 (+) lymph nodes
Treatment: 9 cycles of mFOLFOX
*In remission over 1 year later.

Introduction
- Appendiceal tumors are rare.
- Adenocarcinomas of appendix are only 0.08% of all cancers
- The majority of them are carcinoid but 20% are cystadenocarcinoma (1)(2).
- Cystadenocarcinoma is divided into the mucinous and colonic type. Mucinous type is associated with pseudomyxoma peritonei. (3)

Clinical Presentation of Appendiceal Tumors
Presentation of most tumors is acute appendicitis (49%) and a significant amount are found incidentally during surgery (9.5%).

Diagnosis of Appendiceal Tumors
Benign appendix tumors:
1. Retention cyst.
2. Villous Hyperplasia.
3. Cystadenoma.

Malignant Appendix tumors:
1. mucinous cyst adenocarcinoma
2. Adenocarcinoma
3. Lymphoma
4. Primary Signet Ring Cell Carcinoma
5. Ganglioneuroma

*Diagnosis is rarely made pre-operatively

Treatment of Appendiceal Tumors
- Right hemicolectomy especially when there is involvement of appendiceal or ileocolonic lymph nodes vs simple appendectomy. (4).
- The role of chemotherapy is not well defined, however most oncologists agree on a regimen containing fluorouracil (4).
- Some case reports show remission with the use of FOLFOX with bevacizumab or cetuximab. (1)

Take-Home Points
1. Appendiceal tumors are generally very rare
2. They most commonly present as acute appendicitis
3. Most common subtype is Carcinoid
4. Adenocarcinomas must be managed with at least a right hemicolectomy. Chemotherapy is at times appropriate.

References: