Untreated Metastatic Appendiceal Carcinoid for Five Years
Wallace J. Wang, MD, *Syed Mehdi, MD, *Raina Patel, MD, *Christopher Ashley, MD
Division of Internal Medicine, Albany Medical College and Center and *Stratton VA Medical Center, Albany, NY

Abstract
76 y/o male with no routine medical care presented with abdominal pain and distention for 4 weeks. CT abdomen revealed a 4-6 cm irregularly speculated mesenteric mass within the right lower abdomen with significant infiltration to the adjacent mesentery, circumferentially thickened distal ileum, and numerous peritoneal/omental/mesenteric nodules. He developed a small bowel obstruction where he underwent exploratory laparatomy with appendectomy and small bowel resection. Biopsy of the appendix revealed carcinoid tumor with invasion into the parapendiecal fat, while the small bowel also showed carcinoid tumor with perienteric invasion, mucosal ulceration and sensitivity. His 24 hour urine 5-HIAA levels were elevated at 23.7mg, but as clinically he did not have symptoms of carcinoid syndrome, and treatment with long-acting analogues of somatostatin were not recommended. Over the next 4 years, he remained asymptomatic without wheezing, flushing and diarrhea; with 24 hour urine 5-HIAA steadily increased to peak of 60.6mg. Repeat annual CT scans showed numerous stable peritoneal/omental/mesenteric nodules. He developed a small bowel obstruction where treatment modalities for metastatic carcinoid tumors are orthotopic liver transplant, hepatic artery embolisation, and longacting somatostatin analogues, adjuvant Indium-111, Yttrium-90, Lutetium-177 octreotide-receptor targeted therapy.

Discussion
* Our patient was a 76 y/o male who presented with abdominal pain and distention whereupon CT abdomen revealed an irregularly speculated mesenteric mass and diffuse mesenteric & omental nodules. A laparatomy ensued where an appendectomy was performed and biopsy revealed carcinoid tumor with local invasion. His 24 hour 5-HIAA levels were elevated but he was clinically asymptomatic and closely observed for 5 years without evidence of carcinoid syndrome.
* Carcinoid tumors synthesize serotonin, histamine, prostaglandins and usually arise in the gastrointestinal tract.
* Amongst the various carcinoid tumors, only 10% of those in the small intestine and less than 1% of those in the appendix, develop the syndrome. Normally only 1% of dietary tryptophan is converted to serotonin; however serotonin can be as high as 70% in patients with carcinoid syndrome.
* Episodic flushing of face, neck, upper chest is classic for appendiceal carcinoid. Other symptoms include profuse secretory diarrhea, abdominal cramping, bronchospasm, telangiectasias, and cardiac valvular lesions.
* Symptoms begin abruptly, lasting between 30 seconds and 30 minutes; although prolonged episodes can cause hypotension and cyanosis. Food, alcohol, emotional events, liver palpation and anesthesia can provoke episodes. In the absence of liver metastases, intestinal carcinoid tumors usually do not develop this syndrome.
* Somatostatin analogues provide the most effective symptomatic therapy, although interferon has some utility.

Conclusions
* Overall 5-year survival for carcinoids of the appendix is excellent, around 98%.
* This case illustrates the indolent nature of some appendiceal carcinoids even when they present with metastatic disease.

References: