Duodenal Adenocarcinoma at the Ligament of Treitz: Management and Outcome

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Abstract

Primary small bowel neoplasms at the ligament of Treitz are extremely rare and require advanced surgical technique for extirpation. The insidious onset of disease allows for a delayed presentation, often accompanied by moderate-size growth of the neoplasm, causing intestinal bleeding and bowel obstruction. The partial retroperitoneal location of these tumors pose a unique challenge for surgical resection. We present an unusual case of a primary small bowel adenocarcinoma at the Ligament of Treitz, requiring segmental resection of the fourth portion of the duodenum plus the proximal jejunum.

Keywords

adenocarcinoma, angle or ligament of Trietz, neoplasm

Abbreviations

CT = computerized tomography
EGD = esophagogastroduodenoscopy
FAP = Familial Adenomatous Polyposis
HNPCC = Hereditary non-polyposis colorectal cancer
IHC = immunohistochemistry
MSI = microsatellite instability
PET = positron emission tomography
SUV = standardized uptake value
5-FU = Fluorouracil

Introduction

Although the small bowel constitutes over 75% of the length and 90% of the mucosal surface of the alimentary tract, it is the site of only 1% of gastrointestinal cancers,[1] 1/50th as common as adenocarcinomas of the large intestine,[2] making neoplasms in this location particularly interesting. The mean age at presentation of these tumors is 67 years with higher prevalence in blacks than whites and men than women.[3] The epidemiology of these tumors favors men to women at a rate ratio of 1.5:1.[3] The incidence of carcinomas and carcinoid tumors has been cited as nearly two fold in blacks at 10.6 per million people, compared to 5.6 per million people in whites.[3] Lifestyle factors that are associated with increased risk of small bowel adenocarcinomas include alcohol consumption, ingestion of smoked foods, salt-cured foods, red meats, and refined sugars.[4] Given that most small bowel adenocarcinomas arise from adenomas in the intestinal mucosa, the adenoma–carcinoma sequence well described in the large bowel appears to be recapitulated to some extent in the small bowel, however the exact sequence of genetic changes in the small bowel mucosa has not been well elucidated. Small bowel primary tumors are more frequent in hereditary nonpolyposis colorectal cancer (HNPCC) and familial adenomatous polyposis (FAP) family members, implying similar cancer pathogenesis between these two neoplastic genetic syndromes.[5] Small bowel adenocarcinomas occur with greatest frequency in the duodenum, then decrease in incidence with distal movement throughout the midgut, first jejunum then ileum.[6] This case in particular highlights the rarity of the tumor location at the Ligament of Treitz, thus posing a unique challenge both for identification of the tumor location and for surgical planning: small bowel resection with reestablishment of gastrointestinal continuity.

Case Presentation

A 58-year-old man experiencing slowly progressive lightheadedness, an approximate one-year history of gastrointestinal bleed, iron deficiency anemia, and low-grade upper abdominal vague pain associated with progressive dyspepsia. After a series of negative upper and lower endoscopy studies the patient subsequently underwent a pill endoscopy that showed a proximal small bowel mass lesion. This was followed by a computerized tomography (CT) scan of the abdomen and pelvis which showed a polypoid mass involving the anterior wall of the small intestine, arising from the retroperitoneum at the duodenal-jejunal junction. Internal fluid attenuation of the mass measured 2.6 x 1.8 x 1.7 cm, with additional findings of fat stranding and increased attenuation of the surrounding mesentery. There was no gross regional lymphadenopathy. The patient underwent an endoscopic biopsy of this lesion by esophagogastroduodenoscopy (EGD), which returned with a diagnosis of moderately differentiated adenocarcinoma.

A whole body pre-operative positive emission tomography (PET)—CT was performed, which demonstrated that the neoplasm displayed marked hypermetabolism, showing a standardized uptake value (SUV) max of 19.1, as well as an associated hypermetabolic 1.5 cm lymph node or tumor extension immediately adjacent and anterior to this small bowel segment. The cut off between benign and malignant lesion/nodule is in the SUV range of 2.0-2.5.[7]

The patient was taken for surgery and the small bowel tumor was resected (duodeno-jejunal segmental resection) en bloc with the corresponding jejunal mesentery for wide margins and

nodal clearance (15 nodes harvested). The tumor involved the fourth portion of the duodenum and extended into the proximal jejunum. The duodenal/jejunal mass seemed to be adherent to the mesentery of the transverse colon initially, and by following oncologic principle, no attempt was made initially to separate these two structures (thus avoiding cutting through the tumor extension which could also potentially compromise the mesenteric blood supply to the transverse colon). With division of the transverse colon, our angle of visualization of this small bowel mass in the retroperitoneum vastly improved. The surrounding peritumoral desmoplastic reactions/adhesions were carefully lysed, allowing the mass to be gently dissected (released) off the transverse mesocolon anteriorly and the retroperitoneum posteriorly. Because it was not necessary to resect any transverse colonic mesentery as part of the tumor, we did not compromise the blood supply to the divided colon, thus allowing for easy re-approximation of the transverse colon.

After resection with a 3 cm gross proximal margin at the third portion of the retroperitoneal duodenum and generous resection of the proximal jejunum (8 cm) distally, a hand-sewn side-to-side two layer anastomosis was performed (running 3-0 vicryl full thickness approximation of the small bowel wall with interrupted 4-0 silk Lembert sutures at the serosal layer, and the proximal third portion of duodenum was anastomosed to the free limb of the proximal jejunum (duodenojejunostomy) (Figure 1). The transverse colon was then re-anastomosed in a side-to-side fashion using a 45 mm gastrointestinal anastomosis endoscopic linear cutting stapler, followed by closure of the common enterotomy with two layers in like fashion as the small bowel anastomosis previously described. The mesocolon was then closed with a running 3-0 vicryl suture and then this site was fixed to the relocated proximal jejunum using a 3-0 silk stitch. The neoplasm (Figure 2) was near- circumferential, and centrally eroded into the mucosa, measuring 6.6 x 8.9 x 2.3 cm (axially x circumferentially x radially). It extended 1.1 cm from the anterior muscular wall into free serosal jejunal fat, which was focally retracted near the lesion. No other masses or adenopathy were grossly identified. The pathology report showed that sectioning through the adjacent jejunal fatty mesentery revealed multiple negative lymph nodes (15 total).

Final pathology confirmed that this lesion represented a moderately differentiated adenocarcinoma. Final tumor staging pT 3N0M0G2R0, Stage IIA, with 1.7 cm of proximal free margin, 1.8 cm radial/mesenteric margin and 6.6 cm distal margin. The patient was referred for consideration of adjuvant chemotherapy. He was initially considered for Fluorouracil (5-FU) adjuvant chemotherapy, but it was not offered when his microsatellite instability (MSI) testing of the small bowel tumor revealed that he was MSI-H positive and immunohistochemistry (IHC) negative for mismatch repair gene deficiency.

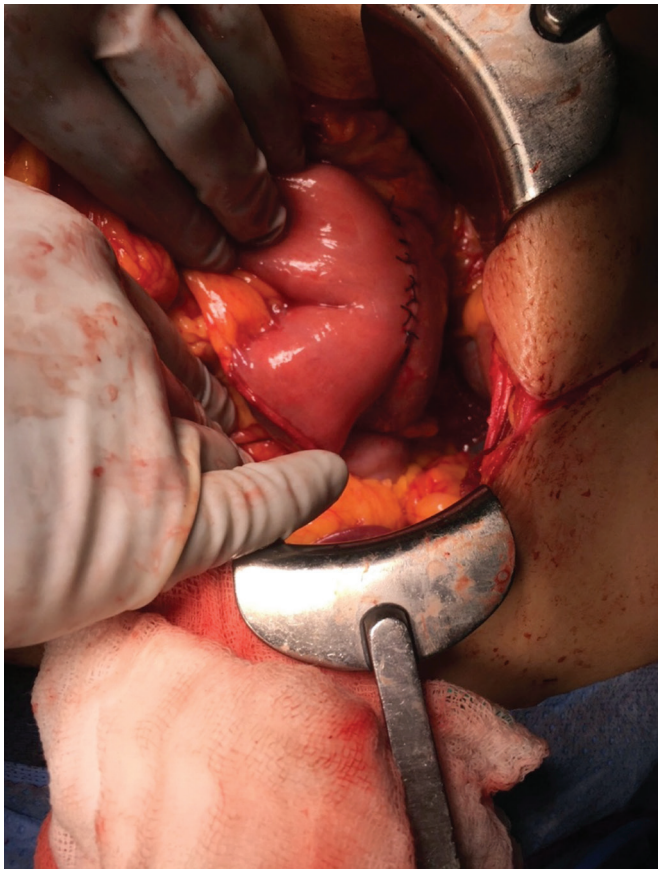

Figure 1. Duodeno-jejunal Anastomosis Following Segmental Resection.

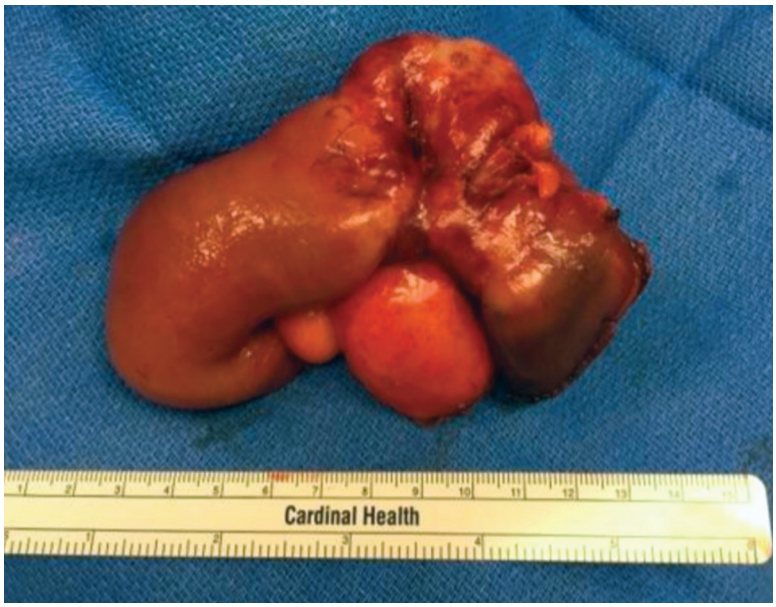

Figure 2. Excised Segment of Small Bowel Containing Neoplasm. Specimen is 16.0 cm in Length and 2.8 cm in Diameter.

The patient has recovered well following his surgery, and does not report any abdominal pain, bloating, or any intestinal bleeding at initial post-op visits to the surgery clinic and on his last visit to the medical oncology clinic he remains asymptomatic with a negative PET/CT 24 months after his surgical procedure. At the most recent follow up, 31 months post surgery, he continues to be asymptomatic without gastrointestinal symptoms: He reports that he is eating well, working regularly and playing golf on weekends.

Discussion

Small bowel malignancies are uncommon, comprising only 1%-2% of gastrointestinal malignancies,[8] and 0.3% of all malignancies.[9] A retrospective study of 1260 patients showed that the most common location of small bowel tumors is the ileum (29.7%), followed by the duodenum (25.3%), and then the jejunum (15.3%) for the entire small intestine. The most prevalent subtype was carcinoid (33%), followed by adenocarcinoma (27%), and lymphoma (16.3%).[9] The prevalence of neoplasms at each location is theorized to be due to the relative concentration gradients of ingested and partially digested carcinogens along the small bowel.[7] The relatively low incidence of neoplasms in the jejunum in particular is thought to be due to increased protection from carcinogen damage and subsequent metaplasia by continuous and rapid turnover of epithelial cells.[9]

Segmental excision of the fourth portion of the duodenum is challenging, as it is a partially retroperitoneal structure, located in a posterior confined space. Of note is the anatomic proximity to the aorta posteriorly, and the stomach superiorly. The reconstructive anastomosis is often unique, and surgical fixation is crucial in order to replicate the functionality of the Ligament of Treitz in suspending the duodenum and preventing gastrointestinal malrotation.

Conclusion

Adenocarcinoma at the Ligament of Treitz is extremely rare. The insidious onset and vague abdominal symptoms, coupled with the difficulty in viewing this location and performing a biopsy on endoscopy, make diagnosing neoplasms at the Ligament of Treitz particularly difficult. Surgical resection for these junctional small bowel adenocarcinomas is also challenging because of the short mesentery and close proximity to the retroperitoneum. We were able to achieve wide surgical margins and yet establish continuity of the upper gastrointestinal tract. Our patient remains asymptomatic and free of disease 31 months after surgery, despite no adjuvant chemotherapy. Studies of follow-up data on a cohort of patients who have undergone excision of similar rare small bowel adenocarcinomas with adjuvant therapy are needed to assess its inherent prognosis and to better evaluate treatment outcomes.

The views expressed in this manuscript are those of the authors and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the US Government.

Conflict of Interest

None of the authors identify any conflict of interest.

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