


A Rare Presentation of Sarcomatoid Carcinoma of Duodenum: A Case Report

Journal of Investigative Medicine High
Impact Case Reports
Volume 10: 1–4
© 2022 American Federation for
Medical Research
DOI: 10.1177/23247096221121392
journals.sagepub.com/home/hic


Jonathan Vincent Reyes, MD¹ , Tasur Seen, MD¹,
Bhanu Singh, MD¹, and Aaron Walfish, MD¹

Abstract

Sarcomatoid tumors are very rare biphasic tumors characterized by a mixture of malignant epithelial and mesenchymal cells that have been usually identified in the lungs with other documented cases in skin, bone, thyroid gland, salivary glands, breast, and genitourinary and gastrointestinal systems. They have an incidence estimated to be 0.5 to 0.8 per 100 000 per year. Three classic features include the presence of a genuine sarcomatous component, no transitional zone between carcinomatous and sarcomatous components, and immunohistochemistry of the sarcomatous component that is positive for mesenchymal markers and negative for epithelial markers. Sarcomatoid carcinoma of the gastrointestinal tract is rare but more commonly found within the stomach, gallbladder, and esophagus. Small bowel involvement is very rare.

Keywords

gastroenterology, hematology oncology

Introduction

The small bowel is a highly active organ utilizing 90% of itself for absorption while comprising 75% of the length of the gastrointestinal tract (GIT).¹ Despite being so active and having high cell turnover, tumors of the small bowel are quite rare and comprise less than 5% of all gastrointestinal (GI) cancers.^{1,2} Dikman and Toker³ characterized sarcomatoid carcinomas in 1973 when it was first coined as enteroblastoma. It has also been called carcinosarcoma, metaplastic carcinoma, spindle cell carcinoma, and pleomorphic carcinoma in the past.⁴ Sarcomatoid carcinoma within the small intestine has an incidence estimated to be 0.5 to 0.8 per 100 000 per year.^{4,6} It affects males more than females with a ratio of 1.5:1.⁷ The average age patients are diagnosed with sarcomatoid carcinoma of the small intestine is 57 years old.⁵ Risk factors have not yet been established; however, few publications cite a possible correlation with long-standing regional enteritis.⁸ Gross imaging appears polypoid or ulcerous fungating mass with or without central ulceration and are frequently necrotic and hemorrhagic with an average of 7 cm in greatest dimension (with a range between 3 and 16 cm).^{4,5,8} Patients most commonly present with anemia as well as nonspecific symptoms including abdominal pain, nausea, vomiting, obstruction, weight loss, GI bleeding, palpable abdominal mass, and fatigue.^{5,8,9}

Case Presentation

A 61-year-old male former smoker with a medical history of hypertension had initially presented to the emergency

department with complaints of nonspecific abdominal pain and approximate 30 lb unintentional weight loss. Apart from his complaints, laboratory values were significant for hemoglobin of 9.5 with a mean corpuscular volume of 94.1. The liver function test on admission was significant for alkaline phosphatase (ALP) of 226, alanine aminotransferase (ALT) of 76, and aspartate aminotransferase (AST) of 71. Total bilirubin was 2.2 with a direct bilirubin of 1.7. The patient remained stable in the emergency room and was discharged with a close follow-up with gastroenterology.

The patient was scheduled for an expedited outpatient endoscopy. The upper endoscopy revealed an edematous pylorus which was traversed with some difficulty as seen in Images 1 and 2; upon passing through the pylorus, there was a medium-sized fungating, polypoid, and ulcerated mass in the anterior wall of the duodenal bulb. The edematous tissue in the region precluded adequate visualization of the lesion, so an over-the-scope cap was used. On close evaluation, the lesion did not appear to invade the pyloric channel, and multiple biopsies were taken. For a

¹Icahn School of Medicine at Mount Sinai Hospital (Elmhurst Hospital Center), New York, NY, USA

Received April 26, 2022. Revised July 29, 2022. Accepted August 7, 2022.

Corresponding Author:

Jonathan Vincent Reyes, MD, Department of Internal Medicine, Icahn School of Medicine at Mount Sinai Hospital (Elmhurst Hospital Center), 79-01 Broadway, New York, NY 11373, USA.

Email: reyesj30@nychhc.org





Image 1. Medium-sized infiltrative, polypoid, and ulcerated mass with stigmata of recent bleeding in the anterior wall of the duodenal bulb as demonstrated through the endoscopic imaging.

comprehensive assessment, a colonoscopy was performed at the same time despite the significant upper endoscopy findings, which were normal. The patient was immediately scheduled for computed tomography (CT) of the abdomen and pelvis, which demonstrated a $7.8 \times 7.0 \times 7.1$ cm irregular, heterogeneous mass which appeared to be contiguous with the duodenum, gastric antrum, and the pancreas. The distal body and tail of the pancreas were atrophic, along with dilatation of the pancreatic duct with a diameter of 6.8 mm. In addition, there were 2 discrete hepatic masses with mild intrahepatic biliary ductal dilatation. The mass also partially encased the celiac axis, right hepatic artery, proximal splenic artery, as well as a segment of the superior mesenteric artery. The main portal vein, splenic vein, and superior mesenteric vein were occluded.

The preliminary results of the biopsy specimens demonstrated a spindle cell neoplasm with surface ulceration. The specimen underwent immunohistochemical staining to rule out other mesenchymal lesions (CK117, S100, CD34, SMA, AE1/AE3), and the specimen was sent to our affiliated tertiary care facility for further review. The final pathologic diagnosis was sarcomatoid carcinoma with osteoclast-like giant cells. The specimen was patchily positive for s100 and negative for MCK, CD117, CD34, and SMA. The spindle cells were focally positive for EMA and negative for AE1/AE3, CAM5.2, and sox10. The patient's case was further discussed in our multidisciplinary tumor board conference. He was determined not to be a surgical candidate, with the proposed treatment plan being palliative chemotherapy. The patient subsequently declined further treatment, instead opting to spend the remainder of his time in comfort with his family.

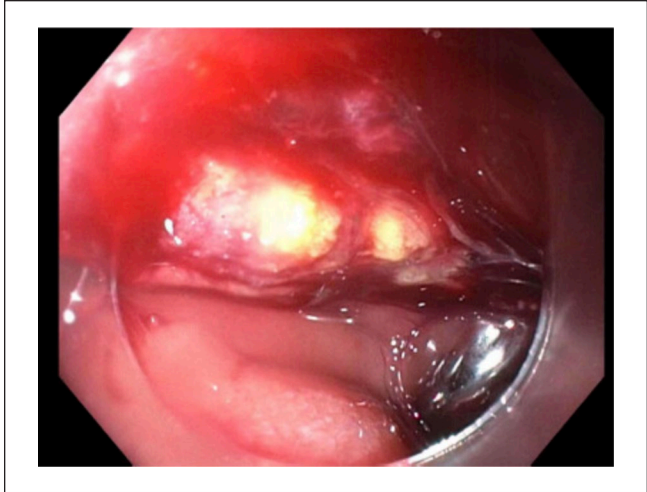


Image 2. Another view of a Medium-sized infiltrative, polypoid, and ulcerated mass with stigmata of recent bleeding in the anterior wall of the duodenal bulb.

Discussion

While tumors within the small intestine are rare, the most common benign tumors are adenoma, leiomyoma, and lipoma and the 4 most common malignant tumors are adenocarcinoma, neuroendocrine tumors, sarcomas, and lymphomas.^{5,10} Although very rare when compared to the most common small intestine tumor, sarcomatoid carcinoma is highly aggressive when compared to adenocarcinoma of the small intestine.² Adenocarcinoma is the most common type of primary small intestinal malignancy accounting for 30% to 50% of all cases and typically presents as early bowel obstruction due to its epithelial origin.^{2,4} Gastrointestinal stromal tumors are the most frequent mesenchymal tumors of the digestive tract, more commonly found in the stomach (40%-70%) and the small bowel (20%-40%) presenting similar to leiomyosarcoma as large intraluminal ulcerating lesions.^{4,6} Melanoma has shown a predisposition to metastasizing to the small bowel.¹⁰ Small bowel tumors also have the propensity to present as an obscure gastrointestinal bleed accounting for the second most common cause, secondary to angiodysplasia.¹⁰ Adenocarcinomas, carcinoids, and lymphomas present as chronic anemia with a gradual more chronic blood loss while gastrointestinal stromal tumors present as an acute anemia with rapid bleeding.¹⁰ To evaluate patients for primary small bowel tumors, video capsule endoscopy and double-balloon enteroscopy are more effective and potentially allow for earlier diagnosis when compared with CT scan as findings are nonspecific and cannot be radiologically distinguished from mesenchymal tumors such as gastrointestinal stromal tumor (GIST), lymphoma, carcinoid, or leiomyosarcoma.⁴⁻⁶

In the literature, sarcomatoid tumors are rare biphasic tumors characterized by a mixture of malignant epithelial and mesenchymal cells that have been identified in multiple

organ systems including skin, bone, thyroid gland, salivary glands, lungs, breast, urinary tract, prostate, liver, pancreas, and GIT.^{4,5,7,11} Three features that are classic for sarcomatoid carcinomas are presence of a genuine sarcomatous component, no transitional zone between carcinomatous and sarcomatous components, and immunohistochemistry of the sarcomatous component that is positive for mesenchymal markers and negative for epithelial markers.¹²

Diagnosis of sarcomatoid carcinoma within the GIT is a very rare finding for this classification of cancer and it is especially rare within the small intestine. Sarcomatoid carcinoma of the GIT is more commonly found within the stomach, gallbladder, and esophagus.⁵ Sarcomatoid carcinomas are rarely found in the small intestine with the most common locations being the ileum and jejunum, while the documented cases of sarcomatoid carcinomas within the duodenum are limited.⁵ Sarcomatoid carcinoma of the small intestine is uncommon and carries a poor prognosis representing a smaller subset of an already rare disease affecting the gastrointestinal system.^{5,13} Typical locations within the upper gastrointestinal system include the stomach, gallbladder, and esophagus.^{4,9} Within the small bowel itself, the cases of sarcomatoid carcinoma typically arise from the jejunum and the ileum, leaving the duodenum an unexpected place.⁴ Sarcomatoid carcinoma in the lower GIT has aggressive clinical courses and often present with signs and symptoms related to distant metastasis with the most common metastatic location being the lungs followed by distant lymph nodes, liver, brain, and pelvic bones.^{4,8} Sarcomatoid carcinoma is a very aggressive cancer that has an increased tendency to metastasize and a low survival rate.⁷ Once diagnosed with sarcomatoid carcinoma, 70% of patients die between 2 and 36 months with an overall 5-year survival rate about 20%.^{8,11} An association, first noted by Han et al, exists between low-expression nuclear protein Ki67 and a relatively favorable prognosis of sarcomatoid carcinoma.⁷ Ki67 is strongly associated with tumor cell proliferation and growth; however, further research is required to confirm this notion.

To achieve an accurate diagnosis of sarcomatoid carcinoma, histology alone is insufficient as small intestine tumors have only minor histological differences and are difficult to distinguish without immunohistochemistry.^{5,7,10} The use of immunohistochemical markers helps the clinician distinguish between small intestine tumors such as epithelioid angiosarcoma, leiomyosarcoma, epithelioid malignant peripheral nerve sheath tumor (MPNST), GIST, lymphoma, and melanoma.⁵ Patients with sarcomatoid carcinoma have immunohistochemical studies that report pleomorphic cells positive for cytokeratin AE1/AE3, CAM 5.2, and vimentin and negative for desmin, muscle-specific actin, CD34, DOG1, c-kit, and S-100.^{2,12,14} In fact, 90% of all reported cases of sarcomatoid carcinomas of the GIT have been positive for vimentin, which is an interfilament protein found in mesenchymal cells.^{7,8} The differential diagnosis between leiomyosarcoma, GIST, angiosarcoma, and schwannoma can

all be determined with immunohistochemistry. If the sample is negative for S-100 and HMB-45, this excludes the diagnosis of schwannoma, epithelioid MPNST, and melanoma.^{2,13} Leiomyosarcomas typically present with positive markers for muscle-specific actin and desmin.^{2,7} Angiosarcoma and GIST typically present with positive markers for C-kit, CD34, and DOG-1, while the inclusion of positive markers for cytokeratin excludes the diagnosis of these 2 cancers.² Other markers that can be used but are not as specific to are epithelial markers including epithelial membrane antigen, carcinoembryonic antigen, and Leu-M1.¹

Currently, the only treatment modality available for patients suffering from sarcomatoid carcinoma is surgery with wide excision of the affected intestine.^{5,7} Chemotherapy, specifically the use of 5-FU and/or cisplatin and/or radiation therapy, has been studied and documented; however, neither has improved survival rates.^{2,7} While the current treatment modalities for sarcomatoid carcinoma are very limited, immunomodulators are being studied and represent hope for the future. Monoclonal antibodies against programmed death protein (PD-1) and its ligand, PD-L1, have shown promising antitumor activity in several malignancies such as lung sarcomatoid carcinoma.⁸ In fact, there is currently a phase II clinical trial researching the use of PD-L1-targeted immunotherapy in patients with lung sarcomatoid carcinoma.¹⁵ The overexpression of PD-L1 noted to originate from the sarcoma component is associated with the presence of tumor-infiltrating macrophages and lymphocytes.¹⁶ Zhang et al⁹ first noted a distinct expression of PD-L1 protein in primary sarcomatoid carcinoma of the jejunum which leads to the possibility of using PD-L1-targeted immunotherapy for sarcomatoid carcinomas of the GIT. Future research is required to study the use of PD-L1 and Ki67 as clinical or prognostic biomarkers for sarcomatoid carcinoma within the small bowel. Clinical trials like the one mentioned previously involving PD-L1-targeted immunotherapy will be closely monitored as successful trials will lead to an additional treatment modality that we can provide our patients.

Conclusion

Sarcomatoid tumors of the small bowel are exceedingly rare. This case report will inform clinicians of this rare diagnosis. Currently, the only treatment modality is surgery with wide excision. Chemotherapy and/or radiation therapy have been studied; however, neither has improved survival rates. Monoclonal antibodies against programmed death protein (PD-1) and its ligand, PD-L1, have shown promising antitumor activity in several malignancies and represent possible future nonsurgical treatment options.

Authors' Note

This case report was presented as an abstract and won a Presidential Award: "A Rare Presentation of Sarcomatoid Carcinoma of

Duodenum: A Case Report” at the American College of Gastroenterology national conference in October 2021.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Verbal informed consent was obtained from the patient for their anonymized information to be published in this article.

ORCID iD

Jonathan Vincent Reyes  <https://orcid.org/0000-0002-2572-6436>

References

- Mittal M, Parikh DA, Jess H, Mann SK. First case of small bowel sarcomatoid carcinoma found by video capsule endoscopy. *Gastroenterology Res.* 2012;5(2):71-73. doi:10.4021/gr414w.
- Lee HM, Cho MS, Kim YI. A surgically resected large sarcomatoid carcinoma of the jejunum: a case report and literature review. *J Gastric Cancer.* 2015;15(2):143-146. doi:10.5230/jgc.2015.15.2.143.
- Dikman SH, Toker C. Enteroblastoma complicating regional enteritis. *Gastroenterology.* 1973;65(3):462-466. doi:10.1016/s0016-5085(19)33079-3.
- Kang KA, Yoon J, Kang G. Sarcomatoid carcinoma of the small intestine: a case report and review of the literature. *J Korean Soc Radiol.* 2013;69(4):295-299. doi:10.3348/jksr.2013.69.4.295.
- Andrawes P, Shariff M, Chang Q, et al. Primary sarcomatoid carcinoma of the small intestine: very rare and aggressive tumour. *BMJ Case Rep.* 2017;2017:bcr2016217895. <https://casereports.bmj.com/content/2017/bcr-2016-217895>. Accessed October 11, 2020.
- Pata F, Sengodan M, Tang CB, et al. Concomitant jejunal sarcomatoid carcinoma and gastric GIST in patient with polymyalgia rheumatica: a case report. *Int J Surg Case Rep.* 2013;4(5):449-452. doi:10.1016/j.ijscr.2013.02.007.
- Han N, Han QH, Liu YZ, Li ZC, Li J. Perforated sarcomatoid carcinoma of the jejunum: case report. *Oncol Lett.* 2013;6(2):562-564. doi:10.3892/ol.2013.1378.
- Kwok CM. Sarcomatoid carcinoma of the jejunum with gastric metastases: a case report and review of the literature. *Int J Surg Case Rep.* 2016;28:161-164. doi:10.1016/j.ijscr.2016.09.046.
- Zhang B, Cheng BO, Wang L, Zhao KE, Zhuo GZ, Ding JH. Primary sarcomatoid carcinoma of the jejunum with massive intra-abdominal hemorrhage: a case report and review of the literature. *Mol Clin Oncol.* 2016;4(5):811-816. doi:10.3892/mco.2016.809.
- Alfonso Puentes N, Jimenez-Alfaro Larrazabal C, García Higuera MI. Sarcomatoid carcinoma of the jejunum presenting as obscure gastrointestinal bleeding in a patient with a history of gliosarcoma. *Gastroenterol Rep (Oxf).* 2014;2(2):150-153. doi:10.1093/gastro/gou007.
- Arshad HS, Dudekula RA, Niazi M, Malik S, Khaja M. A rare case of sarcomatoid carcinoma of the lung with spine metastasis, including a literature review. *Am J Case Rep.* 2017;18:760-765. doi:10.12659/ajcr.904584.
- Izumi H, Yazawa N, Furukawa D, et al. Carcinosarcoma of the ampulla of Vater: a case report and literature review. *Surg Case Rep.* 2016;2(1):102. doi:10.1186/s40792-016-0233-7.
- Lee SE, Park SY. Sarcomatoid carcinoma of the small intestine: a rare and highly aggressive tumor. *J Korean Surg Soc.* 2012;83(5):321-324. doi:10.4174/jkss.2012.83.5.321.
- Yam JL, Rai MP, Nemaikayala DR, et al. Sarcomatoid carcinoma of the duodenum. *BMJ Case Rep.* 2018;2018:bcr-2017-223991. doi:10.1136/bcr-2017-223991.
- Patel S. Nivolumab and Ipilimumab in treating patients with rare tumors—full text view. <https://clinicaltrials.gov/ct2/show/NCT02834013>. Published 2017. Accessed November 26, 2020.
- Li X, Wu D, Liu H, Chen J. Pulmonary sarcomatoid carcinoma: progress, treatment and expectations. *Ther Adv Med Oncol.* 2020;12:1758835920950207. doi:10.1177/1758835920950207.