

Introduction

- Small cell carcinoma most commonly originates in the lung
- Colorectal small cell carcinoma is very rare, comprising 0.2% of all colorectal cancers
- The incidence of small cell lung cancer (SCLC) was found to be 22-times that of extrapulmonary small cell cancer [2]
- Colorectal SCC is highly aggressive and carries a poor prognosis

Case Description

- Patient is a 42-year-old female with a history of GERD and tobacco abuse who presented for rectal pain, 2 episodes of rectal bleeding, and a 2-month history of worsening constipation. She also endorsed night sweats, fatigue, nausea, and poor appetite
- CT abdomen/pelvis showed a perirectal mass measuring 3.3x2.2 cm with adjacent mildly enlarged lymph nodes (Figure A). Colonoscopy then showed an eroded, nodular, and ulcerated mucosa in the distal rectum (Figure B)
- The mass was biopsied and pathology revealed small cell carcinoma (Figure C). Sigmoidoscopy with EUS and rectal biopsy confirmed poorly differentiated small cell carcinoma
- Immunohistochemistry revealed that the tumor cells were positive for CD56 (Figure D), chromogranin, AE1/AE3 and TTF1
- MRI showed T4N2 disease with possible involvement of the left levator muscle in addition to positive suspicious left inguinal lymph nodes
- Patient was started on cisplatin and etoposide therapy

Images



Figure A

CT scan demonstrating perirectal mass measuring 3.3x2.2 cm concerning for metastases with adjacent mildly enlarged lymph nodes.

Figure B

Colonoscopy image showing an eroded, nodular and ulcerated mucosa in the distal rectum



Figure C

H&E staining at 10x magnification demonstrating the rectal tumor adjacent to normal rectal tissue

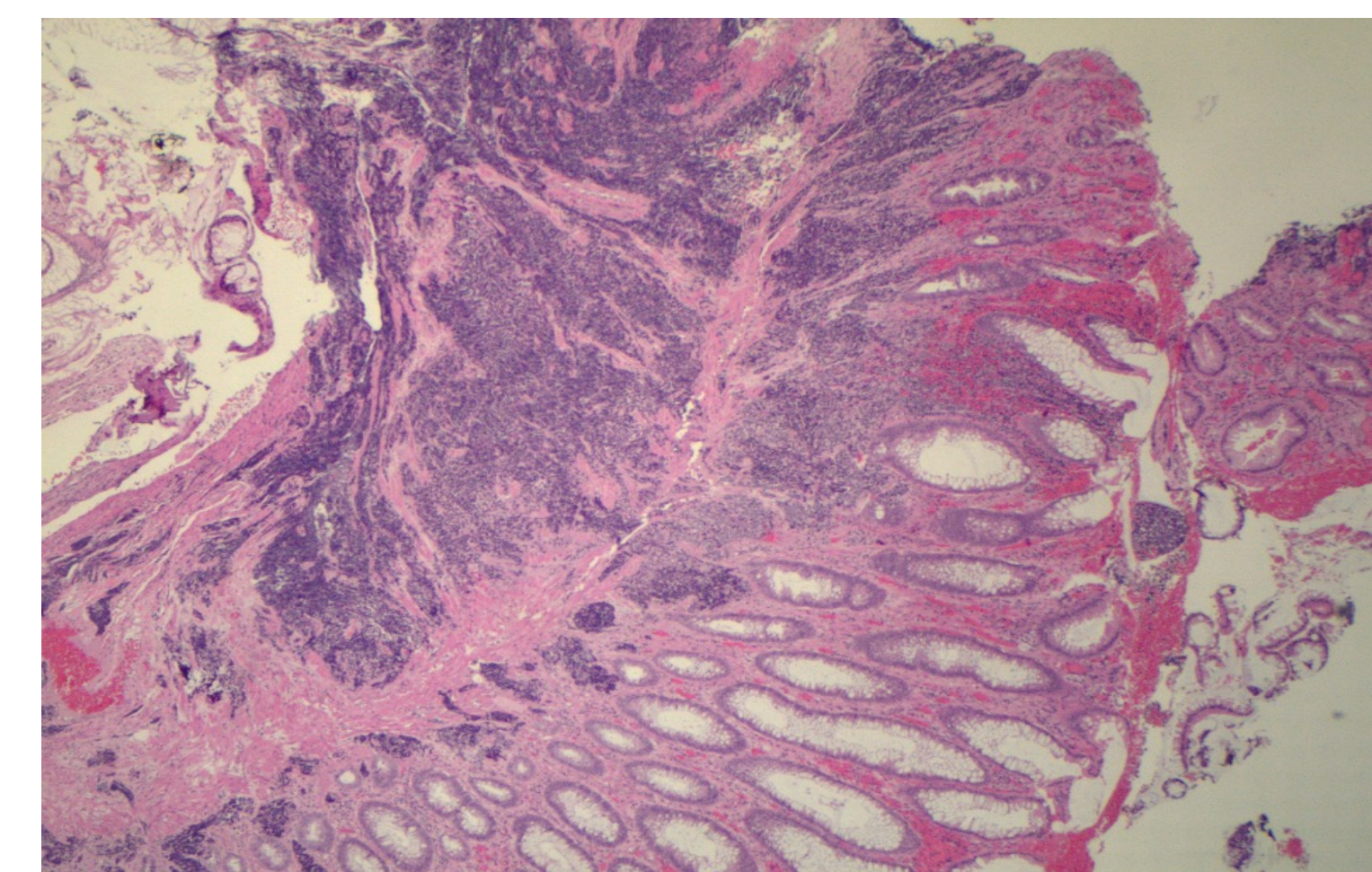
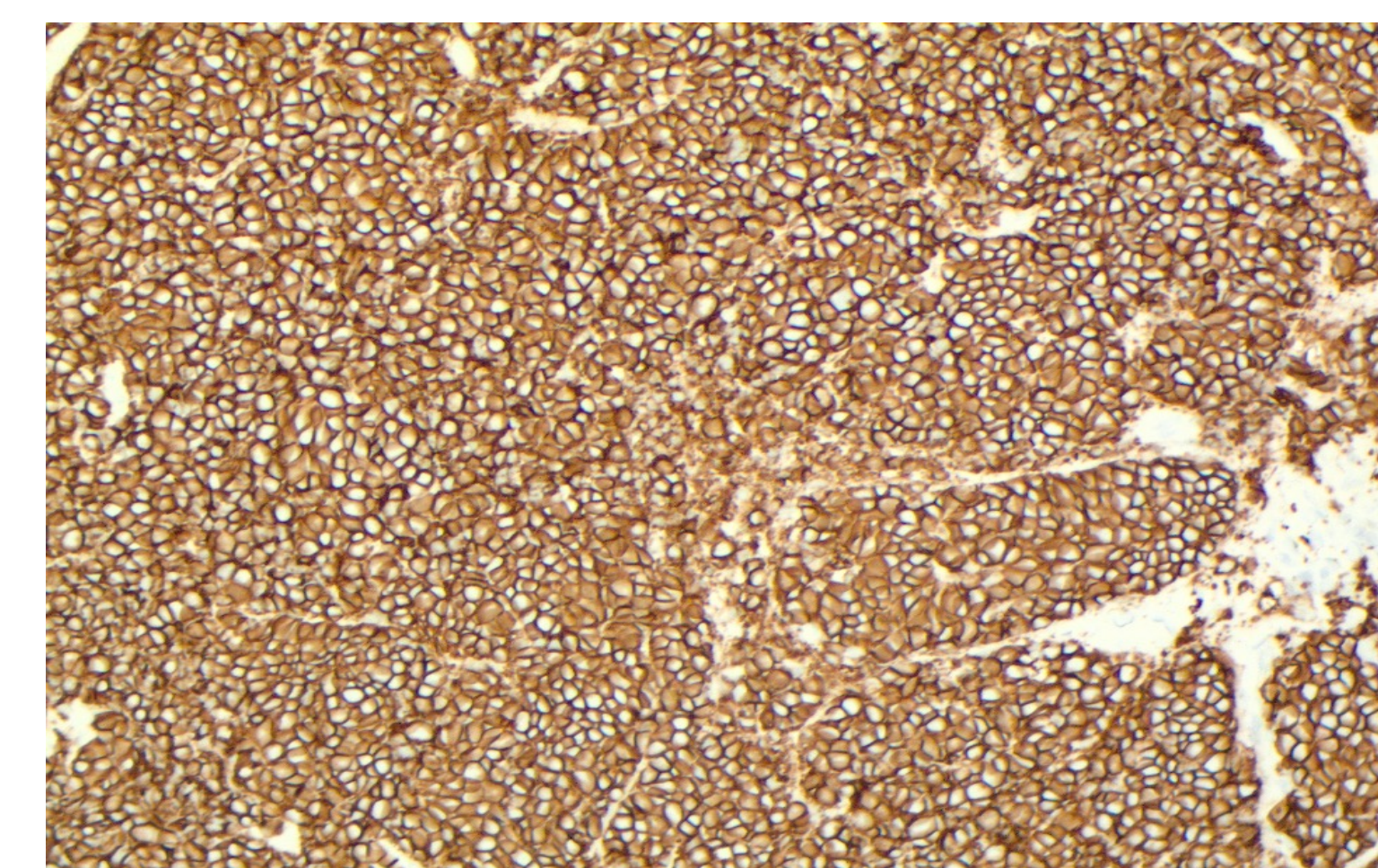


Figure D

Demonstrates Special staining demonstrating positive CD56 tumor cells.



Discussion

- Small cell carcinoma accounts for 0.1% to 1% of all GI malignancies, with the mean age at diagnosis of 60 years old [4]
- Symptoms of rectal small neuroendocrine cancers are similar to those of rectal adenocarcinomas including defecation difficulties, anal discomfort and blood per rectum [3]
- Most patients present with distant metastasis on presentation and have generalized symptoms of malignancy including fatigue, weight loss, and anorexia [1].
- The prognosis of colorectal SCC is generally poor. The rate of lymph node and liver metastases in colorectal SCC patients are 60%-89% and 20%-71%, respectively [5]. Median survival in previous studies was 11 months with palliative chemotherapy and 1 month with best supportive care (BSC) only [4]
- Based on their established role in metastatic SCLC, cisplatin and etoposide have been one of the most widely used regimens in gastroenteropancreatic NEC [4,9], improving median survival to around one year
- Curative surgery is usually attempted in localized disease, although retrospective series indicate that it is rarely curative as a sole therapeutic modality [10]
- Given the high relapse rate observed following radical surgery, platinum-based adjuvant therapy is recommended

References

- [1] Brenner, Baruch, et al. "Small-cell carcinomas of the gastrointestinal tract: a review." *Journal of Clinical Oncology* 22.13 (2004): 2730-2739.
- [2] Doree, Gracia M., et al. "A population-based study of incidence and patient survival of small cell carcinoma in the United States, 1992-2010." *BMC cancer* 15.1 (2015): 1-10.
- [3] Chang S, Choi D, Lee SJ, Lee WJ, Park MH, Kim SW, Lee EK, Jung KT. Neuroendocrine neoplasms of the gastrointestinal tract: classification, pathologic basis, and imaging features. *Radiographics*. 2007;27:1667-1679.
- [4] Sorbye H, Weilin S, Langer SW, Vestermarck LW, Holt N, Osterlund P, Dueland S, Hofslil E, Guren MG, Ohrling K, Birkenmeyer E, Thiss-Evensen E, Blagini M, Gronbaek H, Soveri LM, Olsen IH, Federspiel B, Assmus J, et al. The prognosis of colorectal small cell carcinoma. *Journal of Clinical Oncology* 2014;32:2814-2823.
- [5] Sorbye H, et al. Gastroenteropancreatic high-grade neuroendocrine carcinoma. *Cancer* 2014;120:2814-2823.
- [6] Brenner, L.H., Tang, D.S. Kimura Small-cell carcinomas of the gastrointestinal tract: a review. *J Clin Oncol*. 22 (2004), pp. 2730-2739 doi:10.1200/JCO.2004.09.075
- [7] Rindi G, et al. TNM staging of foregut (neuro) endocrine tumors: a consensus proposal including a grading system. *Virchows Arch* 2006;449:395-401.
- [8] Yamaguchi T, et al. Multicenter retrospective analysis of systemic chemotherapy for advanced neuroendocrine carcinoma of the digestive system. *Cancer Sci* 2014;105:1176-1181.
- [9] Brenner B, et al. Small cell carcinomas of the gastrointestinal tract: clinicopathological features and treatment approach. *Semin Oncol* 2007;34:43-50. 23
- [10] Casas F, et al. Primary small cell carcinoma of the esophagus. *Cancer* 1997;80:1366-1372.
- [12] Beets-Tan RG, Beets GL. Local staging of rectal cancer: a review of imaging. *J Magn Reson Imaging*. 2011 May;33(5):1012-9. doi: 10.1002/jmri.22475. PMID: 21509856.