Anal Neuroendocrine Carcinoma Presenting as Severe Rectal Pain: A Diagnostic and Therapeutic Challenge

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INTRODUCTION

- Neuroendocrine carcinomas (NECs) of the anus and rectum are rare and aggressive.
- Metastatic disease has a poor prognosis, with survival as low as 13.8 months.
- No specific guidelines exist for treating anorectal NEC, treatment follows NCCN recommendations for extrapulmonary NEC
  - Metastatic disease: Chemotherapy.
  - Resectable disease: Resection and/or chemotherapy +/- radiation on a case-by-case basis

CASE REPORT

- We present a 63-year-old female with history of chronic constipation who initially sought medical attention for severe rectal pain and occasional bleeding lasting 2 months.
- Prior colonoscopy, performed 2 years prior, was normal. On digital rectal examination (DRE), the patient experienced significant pain, leading to a diagnosis of anal fissure.
- Non-operative treatment with hydrocortisone was initiated. Due to lack of improvement, she was referred to colorectal surgery.
- An anoscopy, limited by pain, revealed internal hemorrhoids and a palpable nodular area at the posterior midline.
- Further evaluation under anesthesia revealed a **6 cm irregular mass** in the posterior midline, extending towards the right levator muscles.
- Biopsies confirmed **poorly differentiated small cell NEC** with 8 mitoses per high-power field and lymphovascular invasion.
- Carcinoembryonic antigen (CEA) levels were elevated at 8.1 (normal <2.5).
- CT scan of the chest, abdomen, and pelvis identified a **2.9 cm liver lesion** suspicious for metastasis and increased stool burden. MRI showed an **ulcerated lesion at the anorectal junction with exophytic extension involving the internal anal sphincter and abutting the right levator musculature**, along with the liver lesion and multiple subcentimeter bilobar lesions (Figure 1).
- The case was presented at a multidisciplinary tumor board, which recommended systemic chemotherapy followed by restaging and potential resection.

CONCLUSION

- AC, EC, and EP are rare and potentially lethal conditions, with an increased mortality risk when presenting concurrently.
- Given the scarcity of such cases, there is a paucity of clear treatment guidelines, warranting further research.
- Additionally, while the LRINEC score is valuable for assessing necrotizing soft tissue infections, its potential utility in necrotizing/emphysematous intra-abdominal infections should be explored further.

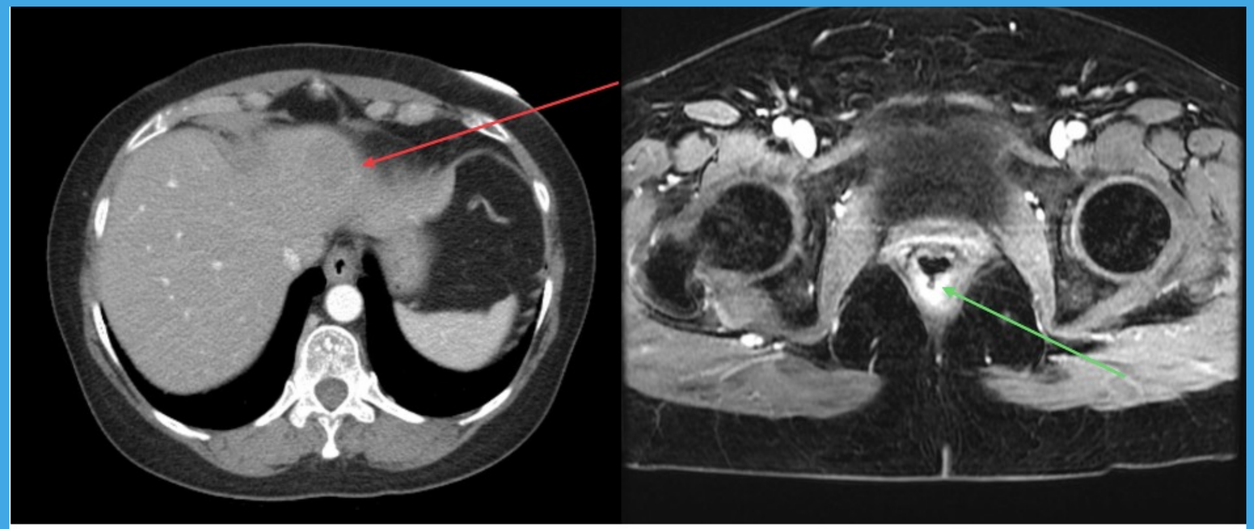


Figure 1: [LEFT]CT image showing 2.9 cm liver lesion (segment two) suspicious for Metastasis (red arrow) and [RIGHT] MRI showing ulcerated lesion at the anorectal junction (green arrow)