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RARE CASE OF A YOUNG MALE WITH METASTATIC MIXED ADENONEUROENDOCRINE TUMOR OF THE ANORECTUM

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Introduction

Modified classification by World Health Organization (WHO) in 2019 has established **Mixed Adenoneuroendocrine Carcinoma** (MANEC) as a subgroup of a broader category of tumors termed **Mixed Neuroendocrine Non-Neuroendocrine Neoplasms** (MiNEN).

MiNEN tumors contain both **neuroendocrine** and **exocrine** tissue representing at least 30% of the tumor volume. Although there remains some debate regarding the aggressiveness of the tumor based on location, MANEC tumors have been confirmed to be a very aggressive entity especially in the presence of a high Ki67 proliferative index and presence of a high-grade neuroendocrine component.

Systematic reviews and population-based studies have identified approximately **800 documented cases** of MANEC involving the **lower gastrointestinal (GI) tract** amongst several additional tumor origin sites.

- Average age of 64 with fairly equal male to female distribution
- Most common site of origin in the lower GI tract is the appendix (60.3%) followed by the colon (29.3%), **rectum (9.2%)** and **anal canal (1.2%)**
- At diagnosis, lymph node positively and metastatic disease have been reported to be 42-49% and 29-67% respectively
- Median overall survival remains poor at **12.3 months** without established treatment guidelines for this inherently aggressive tumor

Conclusion

In this case, we report on a patient with advanced disease who is younger than 98% of the previously reported cases. Additionally, his tumor location within the anorectum is also exceedingly rare and documented in only 10.4% of cases. His tumor also demonstrates a high Ki67 (>90%), high grade endocrine component, lymph node metastasis and liver metastasis which have all been shown to be independent poor prognostic indicators. After discussion at a multidisciplinary tumor board, the patient was started on a combination of Cisplatin and Etoposide with excellent response with no significant residual liver and near complete response in the rectum. Maintenance on Capecitabine planned to be initiated.

Case Presentation

36-year-old male who presented with complaints of intermittent rectal bleeding for several years. Flexible sigmoidoscopy identified a 5.0 cm polyp located 1.5 cm proximal to the anal verge. The patient underwent endoscopic submucosal resection with final pathology revealing a Mixed Adenoneuroendocrine Carcinoma (MANEC) confirmed by histology and immunohistochemistry. Pathology confirmed 1mm negative margins, however the specimen did have positive lymphovascular and perineural invasion. Further staging by CT scan and MRI revealed perirectal, presacral and obturator lymphadenopathy and a PET/CT scan identified suspicious liver lesions that were biopsied and confirmed neuroendocrine metastasis.

Images

Endoscopy (left); **Axial CT scan with lymphadenopathy** (right)





References

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