Excellent Treatment Response of an Aggressive Primary Duodenal Neuroendocrine Carcinoma to Oxaliplatin-Based Chemotherapy

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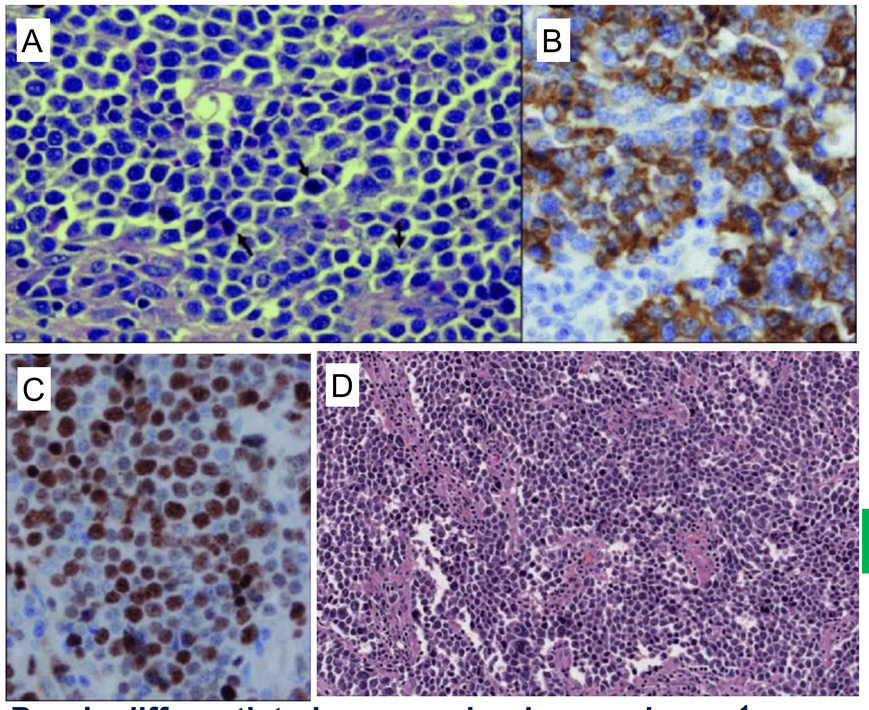
INTRODUCTION

Primary duodenal neuroendocrine carcinoma is a rare and highly aggressive malignancy with very poor prognosis. There is no established treatment due to its rarity. Treatment regimens used for small cell lung cancer are used to treat neuroendocrine carcinoma, due to its clinical and histopathological similarities. Therapeutic strategies are poorly understood and not well defined. There is no standardization of therapy even for early stages, usually a multimodal treatment approach is used. Etoposide based treatment regimens have been used mostly in advanced stages.

CASE DESCRIPTION

A 72-year-old Caucasian male presented to emergency room with abdominal bloating and constipation. He was found to have peritoneal carcinomatosis and marked hydronephrosis in computed tomography of the chest/abdomen/pelvis. He was further evaluated by urology, oncology, and gastroenterology team. He had paracentesis with removal of 4L of ascitic fluid. He underwent esophagogastroduodenoscopy, endoscopic ultrasound, and a colonoscopy for further assessment. He was found to have a large mass in the duodenum which upon biopsy was consistent with grade 4 poorly differentiated neuroendocrine carcinoma. Positron emission tomography dotatate scan, peritoneal biopsy and peritoneal fluid cytology further confirmed metastatic neuroendocrine carcinoma. Tumor markers showed elevated Ca 19-9 and chromogranin levels at presentation.

HISTOLOGY



Poorly differentiated neuroendocrine carcinoma[1]

A. Nuclear pleomorphism and frequent mitotic activity (arrows) **B.** Synaptophysin immunostaining shows strong cytoplasmic positivity. **C.** Ki-67 immunostaining shows a high proliferation index (75% of tumor cells). **D.** Representative patient blood smear showing poorly differentiated neuroendocrine carcinoma

MANAGEMENT

- Started on FOLFOX chemotherapy due to his poor performance status and concerns for intolerability to etoposide
- Completed 12 cycles of chemotherapy with near complete resolution of his disease as evidence by his positron emission tomography Dotatate scan and improvements in his tumor markers

DISCUSSION

- Primary duodenal neuroendocrine carcinoma is a relatively rare malignancy
  - Reported incidence of 0.4-2% among all duodenal malignancies
- Prognosis is poor due to presentation as advanced stage at diagnosis
- Oxaliplatin based regiments have shown to have promising anti-tumor activity in gastrointestinal neuroendocrine cancers
- Available data in duodenal gastrointestinal neuroendocrine cancers are very limited

CONCLUSION

Oxaliplatin based chemotherapy successfully treated an aggressive primary duodenal neuroendocrine carcinoma, adding to the limited literature on treatment of such cancers.

REFERENCES

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