A Rare Presentation of a Duodenal Neuroendocrine Tumor

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INTRODUCTION

- Neuroendocrine tumors (NETs) are rare types of tumors, only contributing to 0.5% of all malignancies. They are predominantly found in females less than 50.
- They are defined as epithelial cells with predominantly neuroendocrine differentiation and consist of a spectrum of tumors emerging from stem cells throughout the body and can occur anywhere in the body.
- Although rare, the incidence over the past few decades has increased. The primary locations for NETs is the gastrointestinal tract (62-67%) and pulmonary tract (22-27%) but duodenal NETs only contribute to 2-3% of all gastrointestinal tract NETs.
- There are 5 unique subtypes of duodenal NETs: duodenal gastrinoma, duodenal somatostatinoma, nonfunctioning duodenal NET, duodenal gangliocytic paraganglioma, and poorly differentiated neuroendocrine carcinomas

CASE PRESENTATION

- A 64-year old female presented to the emergency department secondary to syncope and collapse. During her trauma evaluation, an incidental 3.0 x 3.3 x 3.5 cm lobulated soft tissue mass inferior to the pylorus of the stomach was seen on computed tomography (CT) scan with possible connection to the adjacent bowel.
- EGD showed Grade B esophagitis, external compression of duodenal bulb and diffuse gastritis. AFP, CEA, and CA 19-9 were all negative.
- Surgery team was consulted for resection of the mass and an octreotide scan was performed prior to resection to further evaluate the mass and to check for any signs of metastatic disease.
- The octreotide scan demonstrated intense radiotracer accumulation within the duodenal mass consistent with a neuroendocrine tumor and no other areas of abnormal radiotracer accumulation suspicious for metastatic disease.
- The tumor was found to be attached to the anterior wall of the D2 segment
- Pathology of the resected mass was positive for a well differentiated neuroendocrine tumor with an organoid pattern and homogenous oval-round neoplastic cells with salt-pepper nuclear and pseudoglandular arrangement that was well circumscribed and partially encapsulated with negative margins.
- Immunohistochemistry was positive for AE ⅓, CD56, Synaptophysin, and chromogranin and negative for CD117, DOG-1, CD34 and CD45.

IMAGE 1

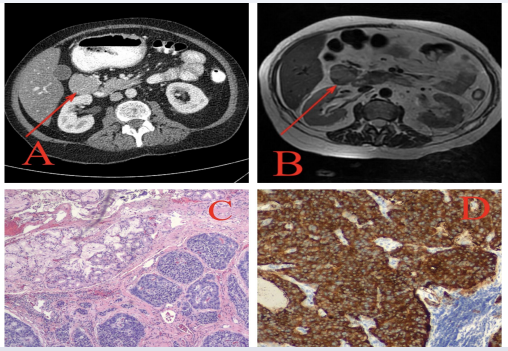

Figure 1: A) CT abdomen and pelvis with contrast displaying a lobulated soft tissue mass inferior to the pylorus measuring approximately 3.0 x 3.3 x 3.5 cm with possible connection with the adjacent bowel (red arrow). B) MRI of the abdomen showing a paraduodenal mass (red arrow). C) Histological slide demonstrating the surface duodenal mucosa and underlying organoid pattern of neuroendocrine tumor. D) An Immunohistochemistry slide demonstrating the positivity of synaptophysin of the neoplastic cells.

DISCUSSION

- The prevalence of NETs has increased over the years due to the improvement in diagnostic tools, such as upper gastrointestinal endoscopy, indicating that prevalence is higher than previously thought.
- The clinical presentation of these neoplasms are similar to other GI tumors, making it hard to clinically detect these type of tumors.
- They are mostly found in the rectum, lungs, small intestines and appendix.
- GI tract accounts for almost 50% of all NETs
- The duodenum is a rare location for such tumors, accounting for only 4% of all GI tumors overall.
- Neuroendocrine tumors are also typically found in those under 50 years old. However, our patient was found to have both a duodenal mass and was over the age of 50 at the time of presentation and diagnosis.
- In fact, the incidence rate in the United States is 0.19 per 100,000. On the other hand, England and Japan have seen a lower prevalence with only 0.04 per 100,000 and 0.17 per 100,000, respectively
- Because almost 90% of the D-NETs are non-functional, the majority of them are incidentally detected on imaging or endoscopy
- While imaging and endoscopy are crucial for detecting them, histopathology is needed to determine the type of tumor.
- The overall prognosis of this disease depends on the classification, tumor type, location, size, staging, and grading. For instance, G1-NET have a very good prognosis. The 5-year survival rate is 80%-85% in people with a well-differentiated D-NET
- To date, a consensus on a conclusive treatment of D-NETs has not been reached but surgical resection seems to be the most radical treatment.
- This case brings to light the importance of further research in diagnosing and treating neuroendocrine tumors and also raises awareness for clinicians to have this in their differential.

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