High Grade Neuroendocrine Tumor of the Gallbladder: A Case Report

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Background

Neuroendocrine tumors (NET) arise from various neuroendocrine cells throughout the body. The most common sites of NETs are the gastrointestinal tract and the respiratory tract, respectively. Of those NETs noted in the gastrointestinal tract, NETs of the gallbladder account for less than 0.3% of these neoplasms. Due to the rarity of NETs of the gallbladder, information regarding the presentation, diagnosis, management, and prognosis of said NETs is sparse. We present a unique case report of a high-grade neuroendocrine carcinoma found in a patient in hopes of providing further insight into this disease process and to allow for further studies elucidating the management and prognosis of NETS of the gallbladder.

Case Presentation

A 53-year-old female with an unremarkable past medical and past surgical history presented with several months of abdominal pain, nausea and vomiting. Work-up with abdominal ultrasound, CT and PET imaging revealed gallstones, a large 4.8 x 4.0 x 3.8 cm loculated mass located centrally within the gallbladder fundus and the presence of porta hepatis lymphadenopathy. The patient underwent endoscopic ultrasound (EUS) and porta hepatis lymph node biopsy which was positive for neoplasm with neuroendocrine features. The patient underwent diagnostic laparoscopy, exploratory laparotomy, core needle biopsy of the gallbladder fundus, pancreatic head mass biopsy, intraoperative ultrasound, and open cholecystectomy. Pathology revealed a high-grade gallbladder neuroendocrine tumor with metastasis to the porta hepatis large lymph nodes and concurrent liver involvement. The patient then underwent post-operative chemoradiation therapy with carboplatin/ etoposide based on recommendations from multi-disciplinary tumor board.

Pre-operative Imaging

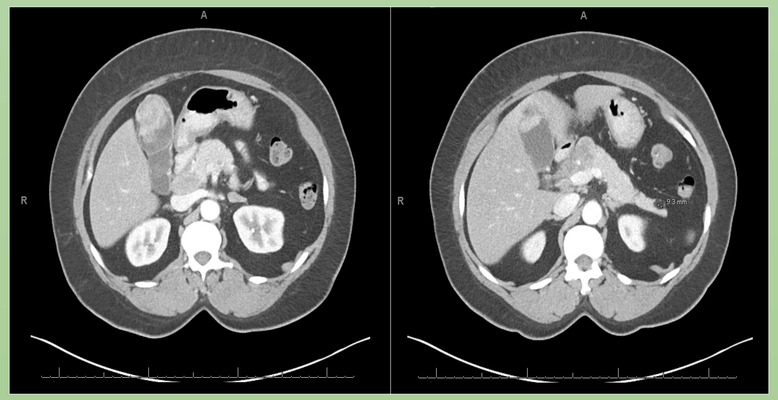


Figure 1 (left): Lobulated enhancing mass centered within the gallbladder fundus which measures approximately 4.0 x 3.8 x 4.8 cm in size.

Figure 2 (right): Periportal/peripancreatic lymphadenopathy. The 3 largest lymph nodes measure 34 x 20 mm, 18 x 16 mm and 31 x 20 mm.

Final Pathology

Peripancreatic lymph nodes fine needle aspiration- Positive for neoplasm with neuroendocrine features. A panel of immunohistochemical stains demonstrated the malignant cells to be positive for chromogranin, synaptophysin, pancytokeratin, and CD56 (weak). Ki67 demonstrated a brisk proliferation index.

Gallbladder, pancreatic head, large lymph nodes- Positive for high grade neuroendocrine tumor with serosal/visceral peritoneal tumor extension and lympho-vascular invasion. No involvement of the cystic duct. Liver involvement was noted with tumor involvement in 3 regional lymph nodes.

Disease Progression

The patient returned to the hospital 1.5 years after initial surgical intervention and completion of adjuvant chemotherapy with progressive complaints of nausea, vomiting, and abdominal distension. CT imaging demonstrated gastric distention secondary to stenosis of the duodenal bulb with evidence of new hepatic metastatic disease in the gallbladder fossa with several nodules adjacent to the duodenum. She ultimately underwent exploratory laparotomy, Roux-en-Y bypass with gastrojejunostomy, jejunojejunostomy, intra-operative ultrasound and duodenal mass/ liver biopsy for gastric outlet obstruction. Pathology revealed grade III neuroendocrine neoplasm. Given progression of her disease process the patient was scheduled to undergo further outpatient chemotherapy with temozolomide/ capecitabine.

Conclusion

Neuroendocrine tumors (NET) of the gallbladder remain a rare neoplasm in the vast array of NETs noted throughout the body. These neoplasms can be managed with a combination of resection and selective post-operative chemoradiation therapy. The key to a favorable prognosis is early detection and multidisciplinary management although further studies must be conducted to provide more concise and systematic recommendations on current practice.

References:

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